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Guest Editorial

J. E. BOONE, M.D., F.R.C.P.(C)

Dr. James Boone was born in Hamilton in 1927 and graduated in Medicine at the University of Toronto in 1951. The following six years were devoted to postgraduate training in Cincinnati, Boston and Toronto. He received his Fellowship in Paediatrics in 1957.

He was in General Practice in West Toronto and Port Credit from 1957 to 1963, when he came on the fulltime paediatric staff of The Hospital for Sick Children with the administrative responsibility of assisting in the direction of the Medical Emergency and Admitting areas.

Dr. Boone's major medical interest is in general paediatrics, but he does have a particular interest in arthritis and rheumatic diseases in children. In 1964, under the auspices of the Canadian Arthritis and Rheumatism Society, he spent two months visiting the Rheumatic Disease Unit dealing particularly with children at Taplow, just outside of London, England.

At present, Dr. Boone is Director of Medical Education at The Hospital for Sick Children and Associate Professor in Paediatrics at the University of Toronto. He is married with a family of three boys. He enjoys the outdoors and the wilderness. In 1969, he paddled 500 miles across the Arctic Barren via the Hanbury and Thelan Rivers to the town of Baker Lake.

Paediatrics is defined as the study, treatment and prevention of disorders of children. It had its beginning in the mid-nineteenth century, when a few Viennese physicians decided to forsake general medicine and begin the study of diseases of infancy. They particularly focused on feeding and nutritional problems, for these were the greatest cause of infant mortality. Their work was well supported by private benefactors, and before long, special clinics and then hospitals were built for children's diseases. However, once admitted to hospital, the children fell prey to infectious epidemics; mortality was high. It became obvious that nutritional and infectious problems must be prevented, for hospital was not the place to anticipate a cure.

The basis of paediatric advances has been prevention. The discovery of vitamins, sterilization, and a scientific basis for formula feeding and intravenous fluids conquered the problems of diarrhea and malnutrition.

While simple bacterial infections can be cured with antibiotics, the greatest stride in infectious disease has been the prevention of polio, measles, tuberculosis, tetanus and diphtheria. Recurrences of rheumatic fever can be prevented with prophylactic Penicillin. Prompt treatment of osteomyelitis and meningitis has prevented life-long disability that was once such a concern in surviving cases.

Tens of different metabolic diseases can be detected through screening programmes. Some, such as phenylketonuria and galactosemia, are successfully managed by dietary manipulation. For most, prevention must be obtained through genetic counselling. For some, sampling of amniotic fluid between the second and third month of pregnancy, will allow detection of homozygous cases in time to prevent the birth of an affected child through therapeutic abortion.

Knowledge of perinatal disease has prevented much of the morbidity and mortality of the neonate. Erythroblastosis is declining through prevention of Rh sensitization. Early therapy has prevented neonatal sepsis in prolonged difficult labour with early rupture of the membranes. Control of temperature and oxygen has prevented cold stress and retrolental fibroplasia. Monitoring blood sugar, calcium and bilirubin in the high risk neonate, has prevented life-long morbidity in many infants. Greater advances in neonatal management will have to include the prevention of the low birth weight delivery, or second best, the delivery of high risk infants adjacent to fully equipped neonatal units.

GUEST EDITORIAL

Challenges to medicine and paediatrics are now coming from outside the profession. In the beginning, medical research and innovation were supported by public generosity. Today, health care is now financed through government legislation. There is a demand for improved efficiency and availability. Health commissions have recognized that the majority of health prevention now carried on in a physician's office could well be administered by others. There has been an unanimous recommendation for the development of a new health worker "physician assistant" who will take on well-defined preventitive tasks, such as well-baby care and immunization. In fact, there is a willingness to see this person make value judgments that will lead to diagnosis and treatment, as long as it is done under the direction and guidance of a licensed physician. Most likely this person will come from the ranks of graduate nurses. Her additional training will have to be a cooperative venture between nursing and medicine.

Paediatrics with its preventative skills must continue to look at the major ailments of children and youth. We are all aware of the present frustrations of our educational system. We have been shown by teachers and parents how a focal intellectual deficit, such as dyslexia, in the otherwise intelligent child can lead to serious learning failures in our present school curriculums. It is believed, that such minor learning problems are responsible for many "drop-outs" and other extremes of adolescent behaviour. Do these deficits arise in the neonatal period? Can they be prevented? Can the emotional overlay that accompanies such problems be treated or prevented? Should we be screening pre-school children for emotional and intellectual deficits, as well as for hearing and vision? The answers to these questions are probably yes. To find the answer will require a close cooperation between family physician, paediatrician, public health nurse, teacher, psychologist and social worker. To prepare physicians for this cooperative role, our undergraduate and graduate curriculums will need to place a greater emphasis on normal behaviour, growth and development, management and educational skills.

Many Thanks

This is the last issue of the JOURNAL in this academic session. Editing this publication for the last two years had many ups and downs. Its viability as an undergraduate medical publication was maintained with the help of a handful of people to whom we express deep gratitude.

May Ho (November 1970), Charles Omole (December 1970) and Richard Simpson (January 1971) devoted appreciable effort to their respective issues. Konrad Godzinski of Maple Leaf Press, made unusual professional effort to ensure smooth publication of this last issue in a race against time.

Wayne Quan (Advertising) and Dave Ward (Circulation) were indispensable again this year. Ross Gillespie managed the budget with remarkable efficiency.

We appreciate the efforts of the authors of the first-rate articles that have blessed the pages of the JOURNAL this year; the articles speak for the writers.

Fred Gentili takes over as Editor-in-Chief with the next issue. As Junior Editor this year, Fred participated fully and admirably in every phase of production of each issue. There is every sign that the quality of this publication will remain at least as high as it ever was. To the class of 7T1, best wishes as each goes a separate way.

T.O.

Behaviour Disorders In Children: Some Perspectives

Laurel J. Dempsey, B.Sc. 7T1

The physician who deals regularly with children in his practice will soon discover that they are as prone to disorders of behaviour as to respiratory tract infections and that the latter are much easier to deal with. This article is designed to give some perspective to the problem, rather than to present symptom clusters in detail. We should consider the physician and the environment, the normal development of the child in several spheres, and finally, the degree and type of abnormal behaviour.

THE SITUATION

A child is brought to the physician with a complaint of disordered behaviour. The first important concept here is that the child has been brought. Most often, the behaviour in question has been a trouble to someone else, and the child is referred because he is troublesome, not necessarily because he is troubled. For the child, the real source of unhappiness may be something else. That is, most behavioural deviations are a symptom of underlying problems, rather than an entity by themselves. To arrive at a solution, if there be one, is often time-consuming and difficult. There are a number of factors which bear on this unwelcome situation and which might help to alleviate the confusion.

These are:

(1) the physician himself;
(2) the culture of the neighbourhood;
(3) the child, by himself, and in the context of his family;
(4) the influence of the school;
(5) special families.

All of these interact in any given behavioural situation.

The physician must first attain some self-understanding. Each of us is a product of a given set of experiences, and although our medical knowledge is reasonably uniform, we do carry prejudices and value systems into our interactions with patients, particularly in the area of behaviour. Both the patient and the physician are subject to the pressures of a swiftly changing culture. The physician must do what he can to enter the "social space" of his patient. This means a thorough study of the prevailing social habits in his practice and of his own frames of reference of "normal behaviour".

In the neighbourhood from which his practice is drawn there may be an easily definable norm for every aspect of behaviour, but increasingly in the large urban areas there are many cultural groups within a small geographical space. Particularly in immigrant populations, the separation of the generations creates two cultural groups, with the parents or grandparents clinging to European values. In some areas, sharp lines are drawn by income levels, with low-rental housing units situated next to middle-income suburbs. Normal behaviour in one group is considered delinquent by the other, and in this day of medical insurance, both may attend the same physician.

The child, the central character in this situation has to be seen as partaking in interactions with his family (the most influence), with other children and adults in the neighbourhood, and with special groups such as schools or clubs. The behaviour disorder may be in one of these spheres, or extend into them all. The physician hopes to establish a relationship with the child, and/or his family. The whole family may have to be treated, since children are a product of their parents child-rearing techniques, and since, except for rare cases, they are all stuck with each other for about twenty years. The family situation has to be assessed, therefore, with respect to parental expectations and the amount of anxiety the behavioural disorder has aroused in the parents.

With the beginning of school, a whole new set of social pressures bear upon the child, and it is at this time that many problems come to attention. Particularly the noisy, aggressive child is in trouble at school. School phobia appears in various disguises, from frank refusal to leave home, to early morning nausea. Various learning disorders and educational retardation are recognised for the first time, as well as some physical complaints. The school constitutes a large sample of "normal" children with whom to compare the patient.

There are special problems of family life in our society, and these include both the very rich and the very poor, the single-parent family, the working mother family, the unmarried mother, and the families which adopt children. Events such as job loss, divorce, death, or geographic relocation may drastically change family constellations. The child himself may have a special gift, or a handicap, to be considered. The task of the physician is to fit these parameters into a harmonic unit for the particular child. Reassurance, teaching, family therapy, drug therapy and the employment of agencies in the community may all be necessary. In addition, there is the element of time. Children are growing, developing organisms and the problem may resolve without active intervention. If the problem is deep, however, the behaviour disorder may be exchanged for another one at another age.

BEHAVIOURAL DEVELOPMENT OF CHILDREN

There are several ways of looking at the development of children. As well as physical development and the achievement of motor skills, children grow psychologically, socially and intellectually.

Freudian Approach

The theory which underlies psychoanalysis is that the child is born with an *id*, a complex of instinctual drives which guides his activity in a direction to obtain satisfaction of his needs. As he grows, external restraints upon his behaviour transform some of his psychic energy into his *ego*, the conscious aspect of personality which guides his activity in keeping with reality. Material with which the ego cannot cope is repressed and lodged in the unconscious. It may come to expression through dreams or gestures, or in mental illness.

The stages of development as seen by Freud are:

The *oral phase*, in which gratification is found in sensations from the mouth. Satisfactory nursing experience is important in this phase of the child's development.

The *anal phase* in which pleasurable sensations arise in the act of defecation, and in which toilet training is a central event. There is the beginning of independence for the child here, for this is a stage in his training in which he may or may not choose to cooperate, possibly setting the stage for a long battle of wills between the child and his parents.

The *phallic phase*, in which, at age two or three, the genitals become the prime centre of gratification. The child's attachment to the parent of the opposite sex is the central phenomenon here.

The *latency phase* from six to twelve in which the child identifies with the parent of the same sex and during which time the *superego* is formed.

These theories were developed during the study of adults with mental illness and are therefore a retrospective way of viewing the development of children. Much insight can be gained however, by their application to deviant behaviour.

The Question of Temperament

Chess[1] has emphasized the role of individual temperament or behaviour style in the study of children, as apart from environmental factors. She has established nine categories of reactivity which can be observed in children from about the age of two to three months.

"(1) . Activity level: The motor component in a child's functioning and the diurnal proportion of active and inactive periods.

(2). Rhythmicity: The regularity and predictability of such functions as hunger, feeding pattern, elimination and sleep-wake cycle.

(3). Approach or withdrawal: The nature of the child's response to a new food, object, or person.

(4). Adaptability: The speed and ease with which current behaviour can be modified in response to altered environmental structuring.

(5). Intensity of Reaction: The energy level of response, irrespective of its quality or direction.

(6). Threshold of responsiveness: The intensity level of stimulation required to evoke a discernable response to sensory stimuli, and social contacts.

(7). Quality of mood: The amount of pleasant joyful or friendly behaviour, as contrasted with unpleasant, unfriendly behaviour or crying.

(8). Distractibility: The effectiveness of extraneous environmental stimuli in interfering with or in altering the direction of the ongoing behaviour.

(9). Attention span and persistence: The length of time a particular activity is pursued, and the continuation of activity in the face of obstacles to maintaining the activity direction."

Using these definitions, clusters or patterns of temperament can be observed even in babies, and the two most striking patterns are of the "easy" and the "difficult" child. The easy child has regular habits, adapts well to new situations, responds positively to stimuli, has mild to moderate reactivity and predominantly positive moods. This child will have fewer behaviour disorders of a disruptive kind, although his very passivity may be a problem in some situations.

On the other extreme, with a high risk of behaviour disturbances, is the difficult child, with irregular habits, nonadaptability, negative responses, intense reactions, bathing, feeding and dressing difficulties and frequent temper tantrums and crying spells. As well, there are other temperamental styles, and it seems that temperament has a large bearing on the child's behavioural development. Techniques of child rearing, such as feeding, can be adjusted to the child's temperament and some of the problems thus circumvented.

Social Development and Role Theory

The discipline of sociology looks at the behaviour of children as a group, and from this study has come theories of the way in which children adapt to their society. The key concepts in this study are *status* and *role*. A status is a position in social structure and to each status is assigned an expected behaviour or role. The stages of socialization are levels of organization of behaviour. The prerequisite for socialization is progressive biological maturation and the acquisition of physical and mental skills. Age confers status on the child according to the society's expectations of children of that age. His position in the family and his family's position in the society also confer a certain status.

The child is socialized by seeing the culture through other people. Those with a close emotional attachment to the child become his *"significant others"*. These include certain members of his core family, other relatives, teachers, and friends. Popular heroes and movie stars may be significant at times. All serve as models by which the child may shape his behaviour. They may also teach directly, rewarding or punishing to achieve desirable behaviour. The most closely involved models have

very strong influences on the child's developing personality, and through them the child is encouraged to internalize the acceptable patterns of behaviour for his status.

Playing at roles is important in socialization. During sessions of "make-believe" the child links certain behaviours with certain roles in society. A girl plays at mothering her doll, thus exploring and defining her expectations of the role of motherhood.

The agencies of socialization in our culture include the family, the school, the peer group and the mass medial.

To begin with, the family has a certain status in the community, defined by ethnic origin, wealth, occupation, religion and other factors. Each child has status within the family, according to age, ability, sex, and health. All the appropriate behaviours for these roles must be learned. The mechanisms of socialization within the family include direct teaching, observation and imitation. An important role to be learned in the home is appropriate sex behaviour.

Next in importance in North American society, and a more organized and uniform agency of socialization is the school. Rules of behaviour are more formalized. The first major conflict for a child may come when the influence of home and school are discordant and he is forced to cope with two roles and standards of behaviour. More often, fortunately, they are in accord and the child's learning is reinforced in school. At this time the child has attained a new status, that of student, and a new independence. During school life, the teacher is the most important significant other.

Peer groups give the child experience in forming relationships with others of equal status, and serve as a testing area for his roles. He becomes more independent and he may be valued in this subgroup of society for talents and abilities which are not valuable elsewhere. He may also be indoctrinated into dyssocial patterns of behaviour if the group becomes a gang.

The phenomenon of mass communication is fairly recent as an agency of socialization. The child is exposed early and often to expression of the popular culture. He learns roles and modes of behaviour from the Western and the detective story which he may internalize. The content of the media is uniform within the society and thus ensures in some part, a uniform understanding of the functions and ways of the society.

Disordered behaviour, as measured against the background of the society may represent a transitory difficulty in making an adjustment of the demands of society, or it may go on to be a permanent antisocial mode of behaviour. The psychopath, for instance, may represent a complete failure of internalization of common values, and the obsessive neurotic a too close adherence to behavioural norms.

Intellectual Development

Intellectual growth in the child depends closely upon the physical maturation of the central nervous system. Intellect can be measured by various tests from about the age of two months. Tests in early infancy give only a rough idea of potential, while those for older children can be scored with reasonable accuracy. There is a strong feeling now that a child's intelligence can be improved by early exposure to learning experiences as long as these are accompanied by adequate mothering. Twin studies have shown a high degree of genetic basis for intelligence. Generally, children with a high degree of intelligence are superior in desirable habits of behaviour to an unselected population, although this is not absolute. Certainly, in our society and our school system, with its emphasis on achievement, a bright child will be more acceptable to both teacher and parent and may have behavioural problems overlooked for this reason. The dull average child in the same school system meets with many frustrations which he may convert into disordered behaviour.

Physical Factors

The child attending for a behaviour disorder should also have a thorough physical examination to rule out physical causes. An example of this is a careful assessment of urinary and bladder function in enuresis. General physical debility such as anemia, fatigue, chronic infection, and malnutrition may exacerbate behavioural symptoms. The cranky, whiny child may simply not be getting enough sleep for his individual needs.

THE CLASSIFICATION OF ABNORMAL BEHAVIOUR

There is not as yet a definitive nomenclature in the area of child psychiatry, but the American Psychiatric Association has produced a diagnostic nomenclature, with ten major sections.[2]

1. Mental Retardation, including that due to gross injury to brain tissue from whatever cause, and that due to psychiatric disturbance and environmental deprivation.
2. Organic Brain Syndromes.
3. Other Psychoses, including juvenile schizophrenia and paranoid states.
4. Neuroses.
5. Personality Disorders.
6. Psychophysiologic Disorders.
7. Special Symptoms which may also occur in other categories including speech and reading disturbance, tics, disorders of sleep and feeding, and enuresis.
8. Transient Situational Disturbances, an acute reaction to environmental stress. This must improve when the stress is removed or the diagnosis must be put in another category.

9. Behaviour Disorders of Childhood and Adolescence: These are more stable than transient disorders and include overactivity, inattentiveness, shyness, over-aggressiveness, timidity and delinquency. Subdivisions of this category include:
   Hyperkinetic reaction (unaccompanied by brain damage)
   Withdrawing reaction (not stable enough to be schizoid)
   Overanxious reaction
   Runaway reaction
   Unsocialized aggressive reaction
   Group delinquent reaction.

10. Non-specific Conditions.

More suitable for organizing our thinking about chlidren who misbehave is the classification proposed by DeCoutere[3].

1. Brain Dysfunction Syndromes
   (a) Cerebral Palsy
   (b) Central blindness, deafness, or receptive sensory loss
   (c) Epilepsy
   (d) Mental Retardation
   (e) Dysphasia
   (f) Specific learning disabilities
   (g) Childhood Psychoses
   (h) Hyperkinetic Syndrome

By brain dysfunction is meant that the cause of the disorder is some insult to brain tissue, not necessarily a demonstrable lesion. It produces dysfunction in the areas of neuromotor skill, sensory reception, regulation of consciousness, cognitive ability, language skill, perception of reality or control of impulsivity. Of particular interest with respect to behaviour disorders is the hyperkinetic syndrome or the syndrome of minimal brain damage. These children have poorly directed activity and they characteristically have a short attention span. They are impulsive and often clumsy, often emotionally labile and socially incompetent. They may or may not have a specific learning disorder.

The second category of DeCoutere, is:

2. Reactive Behaviour Disorders
   (a) Reactive behaviour disorder
   (b) Neurotic behaviour disorder
   (c) Neurotic character disorder

These are changes in behaviour in response to environmental stress, and if they persist they shade into the third category:

3. Neuroses in Childhood:

These are probably less common than was previously thought and include fears, phobias, anxiety, obsessive-compulsive behaviour and hysterical conversion reaction, which is very rare indeed. The fourth category is:

4. Special Symptoms
   (a) Habit disorders: such as thumb-sucking, nose-picking, head-banging, rocking, hair-pulling, and masturbation.
   (b) Enuresis.
   (c) Encopresis.
   (d) Refusal to eat, both on the basis of diminished nutritional requirements in the second year of life and in response to maternal pressures.
   (e) Refusal to sleep.
   (f) Hypochondria, usually with oversolicitous parents and an element of secondary gain.
   (g) Speech Disorders.

5. Emotionality: Disorders of emotional expression and control, including breath-holding attacks, temper tantrums, over-aggressive play with siblings.

In discussing the classification of childhood disorders, another writer, Rutter[4], felt that the following factors should be considered: symptom clusters, response to treatment, long-term prognosis, aetiology, epidemiology, age and sex trends, the presence of concomitant mental retardation, severity, and duration. The prevalence and distribution of emotional and behavioural disorders have been studied and in the next section some of the findings are considered.

PREVALENCE OF BEHAVIOUR DISORDERS

Werry and Quay[5] studied young elementary school children by providing teachers in successive classes with symptom check lists, a common tool for seeking behavioural disorders and one which they feel correlates well with psychiatric examination. They found that the total number of symptoms is more indicative of disturbance than the type of symptom and that boys are more at risk of disordered behaviour than girls. Boys also tended more toward aggressive, acting-out behaviour, whereas girls had more neurotic traits such as shyness and hypersensitivity. All tended to even out as the children reached the upper age limit of the sample (eight). Their checklist included such items as: oddness, bizarre behaviour, restlessness, attention-seeking, staying out late at night, self-consciousness, lacking emotional reactivity, disruptiveness, feelings of inferiority, stealing, shyness, short attention span, lack of self-confidence, inattentiveness, confusion, fighting and laziness.

Rutter and Graham[6] surveyed the entire ten and eleven year old population of the Isle of Wight, using multiple screening procedures, including behaviour questionnaires from both parents and teachers and found that thirteen percent of these youngsters had sufficient disturbances to be referred for further study. Neurotic disorders among girls and antisocial disorders among boys were the most common.

Lapouse and Monk[7] collected 482 children aged six to twelve in Buffalo in a random sample and studied behavioural disorders in the following areas: *Body con-*

trol (posture, falling, clumsiness, speech difficulty, tics, tension phenomena such as nail biting, restlessness). *Behaviour control* (temper loss, wild behaviour, overactivity, disruptiveness). *Combined Habits* (elimination including enuresis, sleep patterns, dreams and nightmares and eating habits). *Behaviour Habits* (sex-appropriate behaviour, compulsive behaviour).

They found that boys had more disturbances of behaviour and that they were of a more antisocial variety. Younger children had more extremes of behaviour, independent of sex. There was not a great difference in pattern between the socio-economic classes represented. "Only children" had a smaller risk of developing behavioural disturbances than children with siblings. In the age group from six to nine, the most frequent disorders were: speech difficulties, tension phenomena, wild behaviour, bedwetting, nightmares, and masturbation. In the nine to twelve year olds the problem seen most frequently were: nailbiting, compulsive behaviour and physical inactivity.

Glidewell, Mensh and Gildea[8] produced a study showing that symptom screening by use of checklists could be used to determine the severity of the disturbance and hence the need for intensive investigation and therapy. The sample consisted of third-grade children and some who had been referred for treatment. They found that the severity of the disturbance correlated with the number of symptoms and that a child with four or more symptoms would be severely disturbed when tested by other techniques.

SPECIAL PROBLEMS

Adolescence

This is a time of great physiological and psychological stress for the developing child. He seeks maturity while coping with a rapidly changing body image, problems of sexual function and identification, decisions regarding his education and increased demands by the school. He may live in domestic tension and experience either parental rejection or overprotection. Many experience school failure for the first time.

Many of the same symptoms indicate disturbance in this age group as in children, but other problems which are introduced at this time are drug addiction and abuse, delinquency, and attempted suicide. One to three percent of all suicides are in adolescents.[9] Other problems are out-of-wedlock pregnancies and sexual promiscuity. Problems with body image, such as obesity or acne are extremely common.

Chronic Illness

Probably the most important factor in the adjustment of a child to a chronic illness is the pre-illness personality. Common initial reactions include regression of behaviour, hostility, anxiety, guilt feelings about the origin of the illness and over-dependence on the parents. Certain conditions such as epilepsy, cystic fibrosis and diabetes mellitus generate strong emotional reactions in both parents and children which require special help and understanding. Various Parents' Associations may be helpful. Strong guilt feelings in the parents of children with inherited disease often cause them to be either rejecting or very over-protective.

Hospitalization

To be in hospital is a traumatic experience especially for the maternal-oriented toddler, to whom no understandable explanation can be given. Among the factors which cause the upset in the child are the unfamiliarity of the hospital, the separation from the parents, and the pre-hospitalization personality. The difficult child who adapts slowly to new situations will have a stormier visit than the placid child. One study[10] showed, however, that a short hospital stay for a general anaesthetic and removal of tonsils and adenoids probably had no lasting effect on the behaviour of the majority of children.

SUMMARY

This article has tried to review some of the background of behaviour disorders in children, omitting specifics of diagnosis and testing, and treatment. Important concepts include the normal processes of socialization and behavioural development, the use of symptom clusters to determine the prevalence of behavioural disturbance, and the necessity for the physician to know himself and his own standards of normal behaviour.

Such understanding can help mitigate the limits which the behaviour patterns of both patient and physician place upon the practice of medicine.

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Cerebral Palsy - A Complex Therapeutic Problem

P. G. Hodgetts 7T1

INTRODUCTION

One often hears of and reads about the concept of a 'medical team' approach to patient assessment and treatment. However, it is extremely seldom when one actually sees this 'team' in action. It is my purpose in this article to discuss cerebral palsy mainly from a therapeutic point of view, pointing up several of the many problem areas, with a slight concentration on the adolescent. It should become apparent that the management of these unfortunate children is indeed one of the best examples of a successful 'team' approach.

GENERAL DESCRIPTION

Cerebral palsy (C.P.) may be defined as a group of non-progressive, persistent disorders resulting from malfunctioning of the motor centres and pathways of the brain, characterized by paralysis, weakness, incoordination or other aberrations of motor function which have their origin prenatally, during birth or before the central nervous system has reached relative maturity. Associated with the motor malfunction is a disturbingly high incidence of seizures, mental retardation, sensory and learning defects, behaviour and emotional disorders. With a prevalence rate of several hundred per 100,000 population, one can see that there are enormous economic, social and medical problems involved in their care.

There are a variety of methods of classification of C.P. including:

by cause (if known) — see below
by type – spastic; athetoid; ataxic; mixed.
by anatomic distribution – hemiplegia; quadriplegia; triplegia, etc.
by functional state and intellectual ability.

Since no one of these systems classifies the C.P. child completely, a combination of all four will provide a total description of the patient and suggest expected problems and prognosis.

In Thelander's study[1] of a group ranging in age from 8 to 25, 60% were males; 47% were spastic quadriplegics; 27% spastic hemiplegics; 9% spastic paraplegics; 8% ataxic; 5% athetoid quadriplegics; 3% rigid quadriplegics; 1% triplegics. To compound the motor disability and the therapeutic problem, he pointed out the incidence of mental retardation: 52% had an I.Q. of less than 50; 13% in the range 50-79; 13% 80-89; 16% 90-109: and only 3% 109 plus. As if this were not problem enough, it is well recognized that there is a high incidence of other physical problems:

Epilepsy: The incidence of grand mal or petit mal fits is as high as 60-80% in some studies in the spastic group, and 12-34% in athetoids. An abnormal EEG was found in 75% of hemiplegics, 58% of whom suffered fits; in 57% of spastics, and 48% of athetoids and mixed groups. There was no relationship between the incidence of epilepsy and the clinical severity of the physical disability. The incidence of fits was lowest in the higher I.Q. ranges, but an abnormal EEG seemed to have no bearing on the I.Q. level.[2]

Ophthalmological problems: These include strabismus, nystagmus, glaucoma, coloboma, visual acuity and field defects, and visual perceptual difficulty in 50-75% of cases.

Hearing loss: This is more common in the athetoid group with an overall incidence of 11-41%.

Speech impairment: This was reported very conservatively in one study at 15%. The figure is no doubt much higher.

Corticosensory and/or kinesthetic loss: Found in about 50%, more commonly in spastic hemiplegics. It should be noted that the lack of sensory stimuli from a limited motor activity, and lack of social stimulation are prime causes of sensory loss.

ETIOLOGY

In the discussion of the therapeutic aspects of any disease, the first stage in treatment, that is, prevention, should be stressed. It would thus be appropriate to briefly look at factors found in reviews of obstetrical and neonatal histories of children with C.P. with the thought of prevention in mind.

It is estimated that approximately one-third of cases of C.P. could be prevented. Combining Thelander's and Anderson's summaries.[1,3] one can see the scope and subtlety of the problem.

MATERNAL DISEASE AND OBSTETRICAL PROBLEMS:

	% of Cases in Which Factor Found
Primigravida	57%
Anemia	12%
Abruptio	4-15%
Vaginal Hemorrhage	12%
Hypertension	14%
Nausea and Vomiting	23%
Toxemia	5-10%
Placenta previa	3%
Trauma	5%
Diabetes	0.8%
Forceps	27%
Breech	4-9%
Version and Extraction	18%
Caesarean Section	3-8%
Precipitate Delivery	5%

INFANT FACTORS:

Twins	5%
Sex – Males	57%
Kernicterus	5%
Neonatal Jaundice	10%
Rh Incompatibility	12%
Cyanosis	30-70%
Required Oxygen	30%
Convulsions	10-30%
Birth Injury and Asphyxia	38-53%
Birth weight:	
over 5½ lbs.	50%
under 5½ lbs.	34%

DIAGNOSIS AND NATURAL HISTORY

SPASTICS

The spasticity is based on a pathologic stretch reflex with increased deep tendon reflexes, clonus, scissoring and contracture development. Early in infancy one may find fixed posturing, opisthotonus, maintained partial tonic neck reflexes, and absent or lateralized stepping and placing reflexes. When placed in the prone position the infant may remain face down.

Hemiplegia: In the upper extremity, the child may ignore his hand, lose sensation and proprioception, and with disuse, growth may decrease, contractures and deformity become established. Aggressive behaviour and seizures are also not uncommon.

Quadriplegia, Diplegia, Paraplegia: In this group contractures and bony deformities, especially subluxed hips, are more frequent. Quadriplegics are often harder to classify accurately since some athetoids have increased reflexes and some spasticity, and spastics may have abnormal movements. In general, if these children don't learn to walk after 6 years, they probably never will. If athetoids delay in developing sitting balance, there will be a longer interval before they learn to walk. Overall, 80% of athetoids will walk, whereas only 70% of spastic quadriplegics will walk. Almost all spastic hemiplegics walk before 3 years.

ATHETOIDS: These children show a picture of involuntary, incoordinate motion, with varying degrees of muscle tension. There may be a history of kernicterus with damage to the basal nuclei. The abnormal movements are usually bilateral and since the lower cranial nerves are affected, there is often accompanying open mouth, drooling, decreased facial expression. It is also noted that during adolescence, quantitative and qualitative changes for the worse take place, especially in spastic diplegics and paraplegics. The reasons for this are unknown.

Denhoff states[3] that between the ages of 6-17 years the mortality rate is over 13 times the normal rate, and that females have a rate 17 times the rate of the general population. It is estimated that given 1000 C.P.'s and 1000 non C.P.'s, 40 years later there would be only 159 C.P.'s alive compared to 933 non C.P.'s.

In many cases early diagnosis may be quite difficult and one should make a thorough attempt to rule out such things as: trauma, poisoning, infections, tumor, diastematomyelia, degenerative disease, Sydenham's chorea, amyotonia congenita, and the muscular dystrophies, before labeling a child with a nonprogressive condition.

In the neonatal period helpful signs include: the presence of feeding problems, irritability, drowsiness, cyanosis, jaundice, respiratory difficulty, abnormal muscle tone, neonatal seizures, persistent Moro and tonic neck reflexes beyond 6-8 months, bruises or cephalohematoma, other developmental anomalies, and tiny, thin, wrinkled infants or large edematous ones.

In infancy, feeding difficulties, poor weight gain and tissue turgor, anemia, and excess motility may be clues. In this age group the abnormal neuromotor development becomes clearer, spasticity appearing before 6 months, athetosis before the end of the first year. One will notice jerky, explosive movements, adducted thighs, hyperreflexia, clonus, mass movement, or in some cases, atonia. In athetoids, purposeless arm and leg movements with straightening of the arms and fist clenching may be present.

In the preschool and older child the diagnosis becomes easier in the more severe cases, and often harder in mild cases. In the latter, the deficit may consist of hyperextension of the fingers and wrist rotation when reaching for items. The child may have an equinovarus deformity at rest, a scissoring gait, or merely poor balance with a widebased gait and alternately clenched and extended fingers. Strabismus and drooling in a hyperactive, destructive or distractible child is often the presenting picture. The ataxic child will have a high-stepping, stumbling, lurching gait usually with nystagmus and tremor; his speech may be monotonous and drawling.

In a study of C.P.'s diagnosed in nursery schools, the presenting complaints were: short attention span in 40%; listless and lethargic in 32%; distractible in 18%; hyperactive in 18%.

TREATMENT

Treatment of the C.P. child must be started early to minimize deprivation and disability. The scope of therapy is so broad and complex that often the treatment of the physical disability is stressed to the point that in later life the patient's psychosocial problems are more crippling. A treatment programme must be established with short and long term goals of a realistic nature based on: auditory, visual, speech, and dental problems; the neurosensory-motor, musculo-skeletal, and nutritional status; hygiene, mobility potential, interpre-

tation to family of patient's role and problems; periodic reassessment and resetting of goals; periodic psychosocial assessment; the keeping of accurate, complete central records to minimize repetition in the child's care; planning for housing needs, educational, recreation and occupational function. One can easily see the number and variety of people from different fields involved in each child's rehabilitation. The physician should assume a central role in the direction of this 'medical team'.

For convenience sake I shall subdivide the treatment principles into: Physical care; Education and Occupation; Recreation and Community care; Social and Psychological care.

PHYSICAL CARE

At an early age (6 mos. - 2 yrs.) the cerebral palsied child should receive passive physiotherapy with a view to developing more normal motor patterns and preventing contractures. Muscle training should be carried out at the child's developmental level. It is important that the parents be carefully instructed so that much of this stage can be carried out at home. The child should be stimulated to reach out to the environment and learn to play with objects just as normal children do. Sensory and perceptual problems should be recognized and treatment programmes planned.

At school age, physiotherapy should continue to help develop the milestones of rolling, sitting, kneeling, squatting, standing and walking. Speech therapy and activities of daily living training should be started early for the purpose of early establishment of relative independence. This is an age of rapid growth predisposing to the development of contractures and deformities. Physio., intelligent bracing, and well-timed and selected surgery are important at this stage. Therapy is often best carried out through games and recreational aids since this best combines enjoyment, normal social interrelationships and therapy. Strength, balance, coordination, and functional posture should be established along with skills in communication and self-help. Again, periodic reassessment and revision of goals is necessary. Personal hygiene should be stressed to establish good habits well before adolescence. Pressure sores, nutritional deficiencies, and dental problems must of course be treated appropriately. The child must always be considered in relation to his family, home and school.

Surgery must be considered for contractures and bony deformities which have not responded to a period of physio. or bracing. The role of orthopedics is to improve muscle balance, posture, mobility, vocational skills and potential. No set rules of indications exist and each case must be assessed carefully and individually. The results of surgery must likewise be compared only to the preoperative status of the child, and the amount of improvement judged on such a basis.

In a study of 605 C.P.'s, each patient underwent an average of 5.8 procedures. Operations on such patients with complex neuromuscular conditions necessitates that careful muscle balance examination and projection of the postoperative effects on this balance, be carried out. To illustrate the importance of orthopedic surgery I will briefly describe some of the commoner procedures and the purpose they serve.[4 5 6]

Procedure	Theoretical Purpose	Results
1. Lengthening middle 1/3 of gastrocnemius aponeurosis with fasciotomy of raphe of soleus.	Improved knee extension and equinus correction without losing push-off	Successful in most
2. Rerouting tendon of Tibialis Post. anterior to malleoli.	Balances dorsi and plantar flexors without losing pronation-supination	Improvement in most
3. Grice subtalar extraarticular arthrodesis	Stabilizes the subtalar joint	Improvement in most
4. Wedge osteotomy of os calcis	Corrects pes planus	Improvement in most
5. Triple arthrodesis	Fusion of the ankle joint in older C.P.'s or ones with poor results of other operations	Well established, good results
6. Egger's procedure (medial hams transferred to femur or adductor magnus insertion; or into patella or quads. tendon at patella	Corrects internal rotation Improves extension of knee	Improvement in most
7. Hamstring tenotomies	Decreases knee flexion deformity and adduction deformity	Not used alone much. Combine with Egger's.
8. Chandler procedure-advancement of patellar tendon for inadequate quads. or high-riding patella	Helps restore knee extension and weight bearing	Helps relieve necessity for bracing in some
9. Hip flexor myotomies	Improve hip flexion deformities	Improvement in most
10. Iliopsoas transfer into post. greater trochanter	Assists hip extension improves support and stability of pelvis in weight bearing	Improves hip extension and lessens gluteal fatigue
11. Adductor myotomy-add. longus ± partial ant. obturator neurectomy	Improve adduction deformity	Improvement in most
12. Brachioradialis transfer for hand deformities: to E.C.R.L. and B. ± fusion of wrist	Corrects flexion deformity at wrist	Depends on selection, intelligence, emotional control, sensation, joint stability
to E.P.L. ± fusion of thumb joints and add. pollicis myotomy	For thumb adducted into palm, with no grasp	
to F.D. Profundus	For adults with severe fixed deformity	Improves finger control so can use as helper hand
13. F.C.U. transplant to E.C.R.B.	Improves wrist in flexion-pronation and with weak extensors	Improvement in most
14. Muscle recession of Pronator teres and F.D.S.	Improves flexion deformity of fingers usually with flexed, pronated wrist.	Decreases strength and straightens fingers.

Again I stress the individualization of surgical therapy; the above procedures are but common examples of what orthopedics has to offer.

EDUCATION AND OCCUPATION

This phase of the C.P.'s treatment often enters the realm of philosophy and it is here that long term goals become most important. Many years of effort on everyone's part, especially the patient's, may be totally wasted

if a potentially capable person is unable to contribute to society in a manner that is personally satisfying. Much stress has always been placed on rehabilitation of the child and adolescent C.P. with a view toward doing some manual work if at all possible. This has in the past led to a relative disregard of the more personal and social side of the patient's well-being. This is probably mainly due to a lack of adequate involvement of the patient himself in setting goals and determining long range plans.

At present it is known that only 20-25% of C.P.'s can reach a state of social and economic independence which might include marriage and children. About 25% will require custodial care, and the remaining 50% will only be relatively independent and require sheltered employment if at all employable.[7] It can be seen that each of these groups will have its own special educational needs and problems. It is suggested that special education programmes should include: leaving the place of residence for some part; concentrating on speech and perceptual disability at an early age; instruction in the use of technical aids eg. typewriters, tape recorders, language boards. The isolation of these children should be minimized by retaining them as much as possible in regular schools. This is of course very difficult to put into practice, raising such problems as: changes in architecture to accommodate a child with braces or a wheelchair; properly trained staff and access of educators to medical consultants; some major changes in public attitudes which would take years to come about.

In 1956 there were 550,000 C.P.'s in the U.S.A. and only 50,000 had the potential for college work.[8] The difficulties involved in obtaining a university degree are many for the C.P. He must be able to meet the physical mobility demands; his educational programme will lag behind at all levels and he must be continually reassured; guidance and counselling services must be more available than at present. In this age, the philosophy of our society dictates that to be a success one must attend university especially if one is to achieve in the occupation market. It is becoming apparent that the value of most university degrees has plummeted, and that despite advanced postgraduate education, jobs which allow one to fully use one's training are scarce. With the present rate of unemployment, how can the cerebral palsied university graduate possibly compete in an open labour market. This seems to put the prime advantage of college experience at a social, personal development level. Perhaps this should be stressed more in counselling these patients. Again one can see the importance of recreation and of developing leisure time activities in the early stages of treatment.

A main problem seems to be that the handicapped are not involved enough in the designing of programmes and facilities. Another problem is that of the nonemployable adolescent. This group seems to virtually disappear once past the paediatric centres' age limit, so that medical follow-up is decreased, programmes are not completed, and the child remains dependent and almost non-functioning in society. He has lost the emotional appeal of a crippled child, and hasn't the economic attraction of the rehabilitated adult. Professional staff also move with less assurance and experience in his treatment programmes. Only a few will gain sheltered employment, which is in itself not the whole answer.

The real limitations to employment seem not to be bound entirely to the disability, but rather to training, self-evaluation, expectations, and the responses of others. In North America we still equate deficiency or failure at work with some kind of moral fault, and this is reflected in the training programmes of C.P.'s. It must be realized that work is not the only value in life, that some C.P.'s should probably not work, at least in our present work ethic society.

RECREATION AND COMMUNITY CARE

For a large number of these children this area of therapy should be stressed. This is also the area requiring the most improvement in facilities available. At an early age through recreation, opportunities for success, achievement, and progress are explored. Physical, social, mental and emotional well being are enhanced, as well as increasing the level of ambulation, dexterity, coordination, cardiovascular and respiratory function.

The community concern is to use recreation activities as a means of assisting the person with a handicap to adjust to the handicap, to the community, to remain in good health and to make use of his leisure time.[9] The problem becomes greater the further from big centres that the child lives. There is a great need here for suitable community facilities, staff, transportation, day care programmes, family counselling, homemaker services, day school programmes.

Another important question is where should the relatively independent C.P. live. Several successful experiments have been done with living in small group homes or apartment complexes, and yet segregation of disabled persons does not seem to be the answer either. It should also be recognized that voluntary bodies can't provide the large programme of community care expected of them. Funds, as always, is the major limiting factor.

PSYCHO-SOCIAL CARE

The world that the C.P. finds himself in today shows an increasing mobility of population towards urban centres with the artificiality and complexity of city life on the increase. The age structure has changed to include more older people. People trained in industry may be retrained 2-3 times, changing their job and often their residence. As economic pressures increase, the space available to the individual will be restricted. Thus the disabled person finds himself in a position with marginal status, increasingly isolated from meaningful

social participation. His situation has been related to that of deviant and oppressed minority groups.[10]

Early in any programme there should be development of interpersonal communication and the beginning of separation from parents and the home. In the adolescent period changes occur which make previous adaptation methods less successful. The child may actually deteriorate physically; he must realize that he won't be cured; his peers will become more acutely aware of differences and he may be left out; he may be more aware of his burden on the family. Certain activities providing social status and feelings of competence may be precluded eg. driving, dancing, athletics, physical attractiveness. Sexual maturation and impulses may be difficult to manage due to poor preparation and education. Leaving school and special treatment centres may be a shock, with the prospects of vocational training, job hunting, changeover in staff, etc. At this time the C.P. must be deeply involved in his own programme and future plans. One must accentuate his assets rather than his disabilities, and make full use of counselling and psychiatric facilities.

Due to his prolonged dependency period the C.P. adolescent will be more frustrated, having had less ability and opportunity for mastering unpleasant situations. He may continue to demand the special treatment he received when younger. The incidence of psychiatric disorders is considered to be higher than in the general population, but there isn't any typical pattern or type.[10]

It should be stressed that the child's arriving at maturity with maximum confidence and ability to compete is as much dependent on his emotional stability as on his motor competence.

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Sickle Cell Anemia

June Hill, 7T1

This disease, at one time a rare entity in Canadian hospitals, is now being seen with increasing frequency due to the influx of immigrants from the Caribbean and Africa. Sickle cell anemia may present in a variety of ways, and its victims may turn up on medical or surgical wards.

DEFINITION

Wintrobe defines sickle cell anemia as "a hereditary and familial form of chronic hemolytic anemia essentially peculiar to Negroes, which is characterized clinically by symptoms of anemia, rheumatoid manifestations, leg ulcers and acute attacks of pain, and is distinguished morphologically by the presence of peculiar sickle shaped and oat shaped red corpuscles, as well as signs of excessive blood destruction and active blood formation."

INHERITANCE AND INCIDENCE

Heterozygotes for the sickling gene are said to possess the sickle cell trait. The homozygous state produces sickle cell anemia.

Sickle cell anemia was first described among Negroes in North America. The incidence of sickle cell trait among U.S. Negroes is 8.5%, incidence of sickle cell anemia varies from 0.3% to 1.3%. Incidence in Central and South America and the West Indies is similar to that in the U.S.

The incidence of the sickling trait in Africa varies widely, highest figures being recorded in Eastern and Central Africa. The incidence was 46% among the pigmoids of East Africa, 18.7% in Nigeria and the Cameroons. There is much evidence to suggest that the sickle cell trait confers a relative immunity to falciparum malaria. It has been proposed that this organism is unable to metabolize HbS as well as normal HbA.

PATHOGENESIS

In sickle cell anemia each erythrocyte contains a high content of abnormal HbS. HbS differs from HbA in that valine is substituted for glutamic acid in the β chain. In environments of low O_2 this abnormal HbS undergoes intramolecular reorganization with change in molecular shape, facilitating "lock and key" interactions between the β chain of one molecule and the α chain of the next. These "lock and key" interactions bring about stacking of HbS molecules within the erythrocyte. Murayama has postulated that molecular stacking is the mechanism responsible for sickling. The sickling impairs blood flow in small vessels leading to obstruction and further anoxia with further sickling.

Sickled erythrocytes have a decreased half life. All symptoms in sickle cell disease are due to (1) anemia or (2) focal vascular occlusion.

CLINICAL MANIFESTATIONS

(a) *Sickle cell trait*

People with sickle cell trait have both HbS and HbA, but more HbA than HbS. Symptoms occur only in situations of severe anoxia, e.g. high altitude flying in nonpressurized aircraft or cyanotic heart disease. Symptoms are due to splenic infarcts, and consist of left upper quadrant pain and tenderness, with occasional splenomegaly and leukocytosis.

Patients with sickle cell trait may also have renal manifestations. The pO_2 in the renal blood supply is lower than the pO_2 in venous blood in other parts of the body. This low pO_2 favours sickling with resultant damage to renal tubular cells. The chief sign is inability to concentrate urine (hyposthenuria). They may occasionally get hematuria.

(b) *Other Heterozygous States*

1. Hb S + HbF

HbF or fetal hemoglobin may be found in combination with HbS. There are few symptoms in this condition because HbF does not react with HbS to produce sickling of the erythrocytes.

2. HbS + HbC

HbC is an abnormal hemoglobin found mainly in Negroes, incidence being highest in West Africa. HbC may exist in combination with HbS, and reacts with HbS to bring about sickling of the erythrocytes. Symptoms in sickle cell-HbC disease are similar to those in homozygous HbS, but milder. There may be mild anemia and splenomegaly, and increased viscosity of the blood may lead to vascular occlusion.

3. HbS + β thalassemia

The clinical features are similar to those in homozygous HbS disease with one major difference. In homozygous HbS the spleen increases in size in infancy, but decreases in size with age due to repeated infarcts. In adult life only fibrotic remains of the spleen persist. In HbS + β thalassemia splenomegaly persists throughout life.

(c) *Clinical Manifestations of Homozygous HbS*

1. Anemic Crisis

An anemic crisis is an episode in which the hemoglobin drops suddenly to low levels due to (1) hemolysis or (2) marrow aplasia. Crises may be precipitated by fatigue, exposure to cold, stress, infections, fever, dehydration and acidosis.

The commonest cause of hemolysis is sequestration of sickled cells in the spleen with their subsequent destruction. This process can produce an acute and profound anemia, occasionally resulting in death. Symptoms are severe pain, fever, and shock. Duration of the crisis is usually 5-7 days.

Transient aplasia of the marrow is occasionally seen in infections. It is a fairly rare cause of anemic crisis.

Treatment is relief of pain with analgesics and plasma expanders such as dextran or fructose. Plasma expanders seem to work by flushing out trapped blood cells.

2. Cardiomegaly

In sickle cell anemia the cardiac output is increased due to chronic anemia, resulting in increased work for the myocardium. Systolic murmurs, accentuation of P_2, and hepatomegaly are found in $>60\%$ of patients. Congestive failure may occur at any time and may be primary, due to failure of the myocardium or secondary to pulmonary complications. Small daily transfusions of packed erythrocytes may bring about decrease in heart size and improvement of cardiac decompensation.

3. Pulmonary Complications

a. Pulmonary thrombosis

Sickling with stagnation and thrombosis may occur in the pulmonary vasculature due to slow flow of blood through the pulmonary arteries and to low mixed venous pO_2. It is sometimes difficult to distinguish between thrombosis and pneumonia, since both may present with chest pain, fever, cough with bloodstreaked sputum and leukocytosis.

b. Respiratory Tract Infections

Respiratory tract infections are common in these patients, perhaps due to their generally debilitated condition. Colds frequently result in pneumonia which classically spreads quickly and disappears slowly. Treatment consists of antibiotics plus daily transfusions of packed erythrocytes (3-5 ml/lb body weight).

4. Renal Complications

The renal complications are hyposthenuria, or inability to concentrate urine due to damage to renal tubular cells, and hematuria. Kidney biopsy in these patients shows glomerular and tubular vessels engorged with sickled erythrocytes. Treatment for hematuria is bedrest, blood transfusion, and steroids.

5. Hepatic Complications

Forty per cent of children with this disease have enlarged tender livers. Low pO_2 in the hepatic circulation causes sickling, and sickled cells block the sinusoids resulting in hepatic cell damage.

Jaundice may occur due to (1) increased destruction of erythrocytes, (2) stone in the bile duct, (3) infection. Fifty per cent of children $>$ 5 years of age with sickle cell anemia have icteric sclerae. Pigment stones in the gallbladder or along the biliary tract are an additional hepatic complication.

6. Skeletal Manifestations

a. Salmonella Osteomyelitis

Children with sickle cell anemia have a high incidence of Salmonella osteomyelitis. Treatment is Chloramphenicol plus Tetracycline or Streptomycin. Surgery may be required to remove necrotic material.

b. Aseptic necrosis

Aseptic necrosis represents infarction of bone resulting from thrombosis of blood vessels supplying that bone. The femoral head is the most common site for aseptic necrosis in this disease. Symptoms may be severe pain or it may be asymptomatic except for the presence of a limp. Aseptic necrosis of the humeral head may also occur, but is less common.

c. Hand-Foot Syndrome

Peak incidence of this syndrome is in the first two years of life. The symptoms are tense swelling and tenderness of the dorsum of the hands and feet, fever, irritability, and occasionally leukocytosis. X-Rays are normal in the acute phase, but X-Rays taken one to two weeks later may show periosteal thickening of metacarpals, metatarsals, and phalanges plus radiolucent areas in these bones. X-Rays eventually revert to normal. The osteolytic changes of Hand-Foot Syndrome are probably due to infarcts in the involved bones.

7. Central Nervous System Manifestations

Symptoms are due either to cerebral anoxia or occlusion of cerebral blood vessels. Symptoms may be transient or permanent and vary from headache, lethargy, weakness to paralysis, convulsions, and coma. The extent and duration of symptoms is a function of the size of the occluded vessels, their areas of supply, and the presence of collateral blood supply.

8. Cutaneous Ulcers

These ulcers occur mostly in teenagers and adults, and are uncommon in young children. The commonest site is on the lower leg, above the ankle. They usually measure 2cm or more in diameter and have sharp margins. Healing takes place over a period of months and results in formation of a thin scar which may break down at any time with reactivation of the ulcer. The ulcers are probably due to thrombosis with occlusion of blood supply to the skin. Treatment consists of control of local infection, bed rest, and transfusions to maintain hemoglobin at normal levels.

9. Inflammatory Soft Tissue Swellings

These swellings appear suddenly around the larger joints — wrist, elbow, knee. The symptoms are reminiscent of rheumatoid arthritis and include pain, swelling, and slight fever lasting 7-10 days. These swellings are probably a tissue reaction to local vascular occlusion. Indomethacin has been found effective in treatment of this phenomenon.

10. Priapism

Priapism is a painful penile erection which may persist for one or two weeks. It chiefly affects children between the ages of five and thirteen, and in sickle cell disease is due to thrombosis of the vascular spaces of the corpora cavernosa. Treatment is aspiration of corpora cavernosa.

11. Retarded Physical Growth and Development

Physical development is retarded in these individuals due to chronic anemia and anoxia of all tissues. They are frequently of short stature, in poor nutritional state, and may be hypogonad. The appearance of the secondary sex characteristics and onset of menarche are often delayed.

HEMATOLOGIC PICTURE IN SICKLE CELL ANEMIA

1. Hemoglobin – reduced
2. Stained smear of peripheral blood – shows mostly round or oval erythrocytes, a few elongated or oval erythrocytes with round or pointed ends, many target cells, occasional nucleated erythrocytes.

   The degree of sickling on a peripheral smear may be minor. A better way to demonstrate sickling is to seal a drop of blood under a coverslip and leave it for two to six hours. If at the end of this time the cells have become long and pointed, the smear is positive.
3. Bone marrow – Fifty to seventy per cent of the marrow is occupied by nucleated erythrocytes, mostly orthochromatic normoblasts. There may be many eosinophils, and there may be a "shift to the left" in the myeloid series.
4. Reticulocyte count – increased (5%-25%).
5. Platelet count – increased (300,000-500,000).
6. White blood count and differential
   – elevated (10,000-30,000) during active hemolysis;
   – there may also be an eosinophilia.
7. Hemoglobin electrophoresis – HbS moves toward the anode more slowly than HbA.

DIAGNOSIS

The clinical features of sickle cell anemia are so varied and mimic so many disorders, that if the possibility of this disease is not kept in mind diagnosis may be missed. Criteria for diagnosis are anemia with high reticulocyte count, sickling of red cells on wet prep, leukocytosis, and elevated serum bilirubin.

COURSE & PROGNOSIS

These patients have a chronic anemia and jaundice with occasional painful anemic crises and other aforementioned complications. The outcome is ultimately fatal, and most patients die in the first decade. A few survive to adulthood with good care.

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TO BE HELD IN TORONTO, MAY 1971

The Canadian Hepatic Foundation which provides support for research and education in the causes, diagnosis, prevention and treatment of diseases of the liver, will conduct its first scientific meeting, a "Symposium on Viral Hepatitis", in Toronto, on May 28-39, 1971.

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Enquiries should be directed to: Carl T. Grant, Secretary, The Canadian Hepatic Foundation, 199 Bay Street, TORONTO 1, Ontario.

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Mycoplasma In Human Disease

Joseph Greenbaum, B.Sc. 7T1

INTRODUCTION

Every micro-organism has its characteristic spectrum of pathogenicity. For the vast majority of micro-organisms, these spectra are now well defined. Thus, Shigella, infects only the G.I. tract; Pneumococcus multiplies in the respiratory tract and occasionally the meninges; Streptococcus has a wider range invading the respiratory tract, the skin, the female genital tract and rarely the blood stream. For the most part these clinical patterns of disease are basic knowledge in microbiology. Mycoplasma are an exception. Although the first member of the group was identified during the classical period of bacteriology in the late nineteenth century, only one species of Mycoplasma has so far been unequivocally proven to be a human pathogen. In 1962 it was finally shown that the Eaton agent causing primary atypical pneumonia was a Mycoplasma. It was named *Mycoplasma pneumoniae*. Controversy still rages over the role of Mycoplasma in other such varied "infections" as non-gonococcal urethritis, Stevens-Johnson Syndrome, rheumatoid arthritis, septic abortion, and infectious polyneuritis (Guillain-Barré Syndrome). Thus a precise pattern of Mycoplasma infection remains elusive.

MORPHOLOGY AND CLASSIFICATION

Human Mycoplasma comprise less than 10 species excluding the T strains. Forming characteristic microscopic "fried egg-like" colonies on artificial medium, they are quite distinctive in being small (100 - 300 mu) organisms without cell walls. Instead, Mycoplasma are bound by a triple layered plasma membrane which may be of great importance in the causation of disease as we shall see later. Their final shapes under the electron microscope are varied, the pleomorphism depending on the fixation and various other factors. Mycoplasma size falls within the range of some of the larger viruses; however they contain both DNA and RNA and possess independent metabolic systems and thus are not viruses. Whether Mycoplasma have any relationship to higher bacteria is controversial; it is clear however that they must be differentiated from the L phase of bacteria. (L stands for Lister Institute where these forms were first discovered.) L phase bacteria are produced under the influence of substances interfering with bacterial cell wall synthesis (eg. penicillin). L phase bacteria and Mycoplasma have been shown to be genetically quite distinct. However they share many physical properties — both lack a solid cell wall and both multiply by the formation of small elements (elementary bodies). There is some doubt as to whether the formation of L phase bacteria is pathogenic, there being recent reports of the isolation of L phase variants from patients with a variety of surgical conditions, with apthous stomatitis, septicemia and chronic infections.

Mycoplasma T strains (so called because of the "tiny" colonies formed) are organisms of about 300 mu diameter, which reproduce primarily by means of a multidirectional budding process to produce ramifying elements. These continue to multiply, forming a "growth flock" or colony.

The artificial media that support growth of Mycoplasma must be highly enriched with serum and yeast extract – even then growth is very slow, 1-3 weeks being required for isolation. Measurement of specific antibodies can be performed by such serological techniques as immunofluorescence (IMF), the complement fixation (CF), the tetrazolium reduction inhibition (TRI), and the fermentation or growth inhibition (FI). For routine purposes the CF test is the most simple and reliable, but is the least sensitive. The others are highly sensitive but laborious and time consuming. Clinically CF Antibodies, in contrast to IMF Antibodies, remain elevated for a long time.

RESPIRATORY TRACT INFECTION

Classically human infection with *Mycoplasma pneumoniae* is manifested as pneumonia. The incubation period is 2-3 weeks and the onset is usually insidious. Symptoms are generalized and the illness may mimic any other acute respiratory disease such as influenza. Usually the severity of symptoms is less than the physical signs would indicate. A dry cough is not uncommon. Rales are prominent together with dullness to percussion. X-ray usually shows involvement of the lower lobes and the type of pneumonia is described as interstitial or broncopneumonic. However inspection and auscultation may be quite normal despite an extensive infiltrate on X-ray. The ESR may be as high as 80 and the WBC may be normal or moderately elevated. Differential diagnosis includes respiratory syncytial and parainfluenza virus infection in children under five; adenovirus pneumonia, Q fever and psittacosis at any age; histoplasmosis and coccidioidomycosis in various geographical locations; and occasionally TB when the X-ray infiltrate is in the upper lobes.

TABLE 1:

CLINICAL APPROACH TO DIAGNOSIS OF *MYCOPLASMA PNEUMONIAE* PNEUMONIA

I. Differentiation of bacterial from nonbacterial pneumonia[a]
   A. Findings that suggest nonbacterial etiology
      1. Presence of marked headache, lassitude, and/or myalgias.
      2. Definite nasopharyngeal disease.
      3. Nonproductive cough and tenderness of the trachea.

4. Peripheral leucocyte count less than 15,000 per mm^3.
5. More pulmonary involvement on chest X-ray than is suggested by findings on physical examination of the chest.
6. Lack of a predominant organism on a gram stain of sputum.

B. Findings that suggest bacterial etiology
1. Conditions associated with bacterial pneumonia, e.g., alcoholism, chronic pulmonary disease.
2. Occurrence of shaking chills.
3. Presence of marked pleuritic pain and grunting respirations.
4. Presence of purulent and/or bloody sputum.
5. Presence of potential bacterial pathogen as the predominant organism in smear or culture of sputum.
6. Peripheral leucocyte count greater than 15,000, especially if there is pronounced neutrophilia (>90 percent).

II. Differentiation of *M. pneumoniae* pneumonia from other causes of nonbacterial pneumonia.
A. Findings that suggest *M. pneumoniae* etiology
1. Age 5-30 years.
2. Occurrence in summer or fall (because frequency of other causes is low at this time; occurrence in spring and winter also compatible).
3. History of exposure to person with respiratory illness 3 weeks before onset.
4. Insidious onset.
5. Presence of myringitis, particularly if bleb or bullae are present.

B. Findings that suggest other causes of nonbacterial pneumonia, e.g., influenza, adenovirus, psittacosis, Q fever
1. Exposure to parakeets, ducks, and other avian species known to carry the psittacosis agent.
2. Exposure to livestock or other known source of Q fever.
3. Presence of epidemic influenza or other virus infection.

III. Confirmatory findings for *M. pneumoniae* pneumonia
A. Presumptive
1. Rise of serum cold hemagglutinins (about 50 percent of cases).
2. Rise of serum Streptococcus MG agglutinins (about 30 percent of cases).

B. Definite
1. Isolation of organism.
2. Rise in specific serum antibody.

[a] Appearance of the chest X-ray is of little aid.
Reproduced from L. Hayflick, The Mycoplasmatales and the L phase of Bacteria.

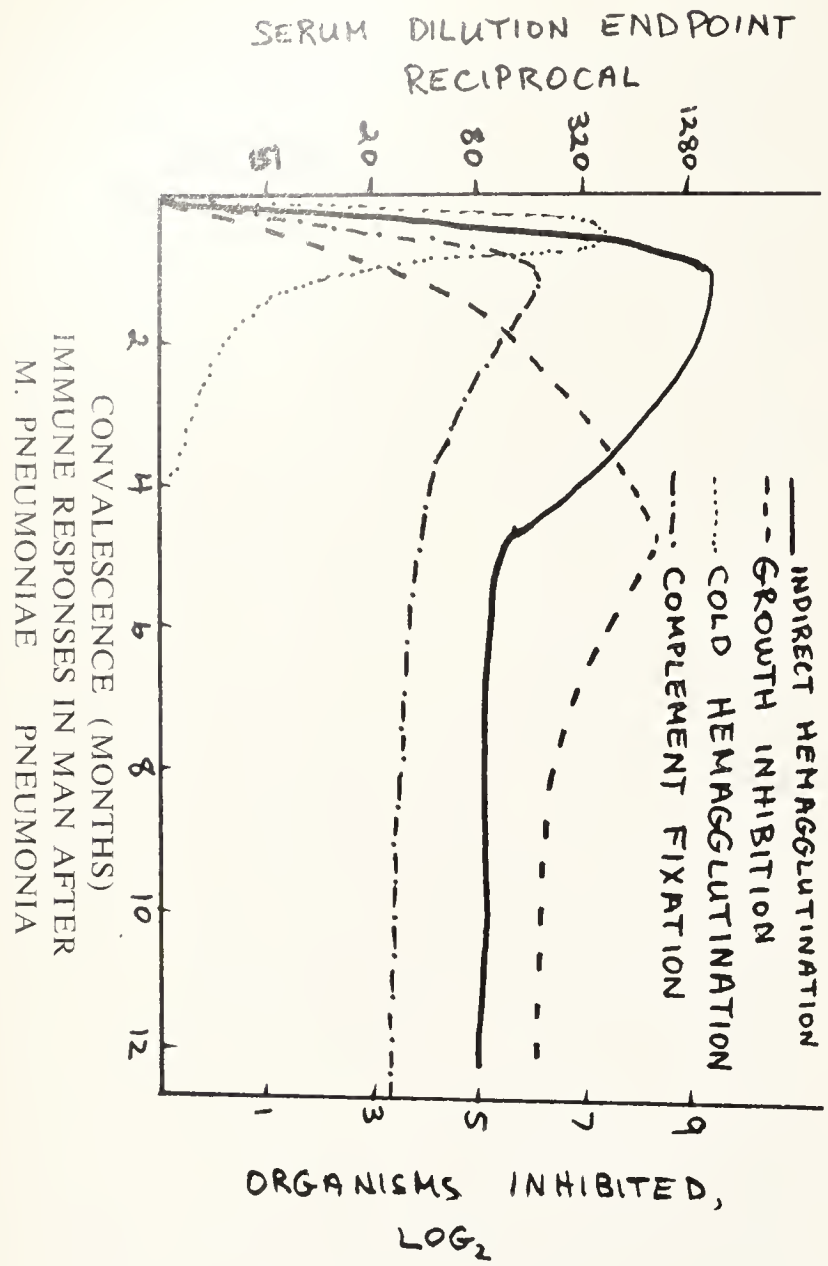
Symptomatology in those infected ranges from pneumonia, bronchitis, laryngotracheobronchitis, and upper respiratory infection to asymptomatic infection. As with pneumonia due to *M. pneumoniae,* there are no clinical characteristics of the nonpneumonic infections which allow their easy identification. Usually the course of the disease is uncomplicated and self limited, although prolonged – slow recovery with malaise, weakness and persistent cough being pronounced, especially in adults. Serious complications are rare. Some of the more common complications include bullous hemorrhagic myringitis, otitis media and skin rashes – these are of some significance. Bacterial superinfection is uncommon. Rare complications include hemolytic anemia with a positive direct Coombs test, thrombocytopenia, coagulation defects, encephalitis and aseptic meningitis.

Several serological responses may occur in *Mycoplasma pneumoniae* infection. A rapid and short-lived rise in the cold haemagglutinin titre may occur but this is non specific since it may be positive in adenoviral and other pneumonias. The height of the cold agglutinin titre is directly related to the severity of the pneumonic involvement. Other serological non-specific responses include biological false-positive tests for syphilis and complement fixing antibodies to lung tissue. The specific responses have been mentioned above. Further serology will be mentioned later. Rarely reinfections may occur in those having specific circulating antibody to Mycoplasma – but these are generally milder than infections in non-immune people.

Epidemiological studies have shown that *Mycoplasma pneumoniae* infections occur throughout the world in such disparate locations as Europe, Asia, the Middle East and the Far East. Infection occurs in all months of the year, with a peak incidence in late summer or early fall. *Mycoplasma pneumoniae* is endemic in many North American metropolitan centres, but a periodicity in the incidence of infection is found, epidemics occuring in cycles of perhaps 5-6 years. The epidemic wave may take months to peak, probably because of the long incubation period and low communicability of the disease. There have also been localized outbreaks in families, schools, military installations and cities.

The highest rate of *Mycoplasma pneumoniae* pneumonia is in the 5-9 year age group; the second highest is in children 10-14 years old. In the 15-19 age group the absolute rates are much lower, but because the overall incidence of pneumonia is so low in teenagers, the incidence of infection due to M. pneumoniae is proportionately highest. This incidence is in contrast to the tendency of respiratory viruses to infect the very young, and pneumococcus the very young and the very old.

This lower frequency of *Mycoplasma pneumoniae* pneumonia in children under 5 can be explained in two ways: first – the milder symptoms allows infection to



go unrecognized more frequently; second – the organism requires close contact for transmission so that adequate exposure does not occur unless the child has older siblings or is old enough to acquire the infection from contact with peers.

These characteristics of *Mycoplasma pneumoniae* – first, its long incubation period and second, the close association necessary for infection, cause the spread of the organism to be very slow. In one small community study, it took three months for the infection to spread among four households; eventually however most susceptible contacts become infected. Another important factor in *Mycoplasma pneumoniae* epidemiology is its prolonged carrier state which can last up to 5 months. In summary then, an infection of this type cannot be expected to occur explosively in epidemics such as those caused by influenza; rather, endemic spread can be anticipated. Community spread in indolent epidemics will take place predominantly among the susceptible child population.

What is the role of other Mycoplasma species in disease? The pathogenicity of *Mycoplasma pneumoniae* is no longer doubted; but the place of other Mycoplasma strains is still hotly debated, and many contradictory statements will be found on this subject in recent medical literature. Much of what follows is only one interpretation of the evidence.

GENITOURINARY INFECTIONS

The organisms incriminated in the causation of non-gonococcal urethritis (NGU) includes *E. coli, Strep. faecalis, Staph. aureus, Haemophilus vaginalis, Trichomonas vaginalis* and several fungal species. Studies trying to implicate *Mycoplasma hominis* type 1 in this venereal disease are unconvincing; *Mycoplasma hominis* is isolated both in a large part of the normal sexually active population and in a majority of patients with NGU. *Mycoplasma hominis* has been found to be resistant to erythromycin whereas NGU responds well to this antibiotic. *Mycoplasma hominis* is thus probably a normal, non-pathogenic commensal in the urinary tract.

The evidence linking T strain Mycoplasmas to non gonococcal urethritis is much more convincing. One study showed that T strains were present in up to 90% of cases of NGU in males, whereas their natural occurence in the genitourinary tract of normal male controls of similar age, occupation and sexual activity generally is under 25%. Similar studies showed that T strain NGU could be successfully eradicated with erythromycin and then re-introduced by the presence of T strains in the sex partner; moreover simultaneous treatment of both partners broke the "ping-pong" treatment-reinfection cycle and at the same time T strains disappeared from both individuals. The fact that T strains can be found in the absence of clinical disease may indicate merely that these strains can, like many bacteria remain latent in the host. The gonococcus for example is frequently (up to 57% in one study) isolated from the genital tract in asymptomatic individuals. The question remains to be answered, however, whether T strains in normal controls are non pathogenic strains or potentially pathogenic strains dormant until triggered into activity by as yet unknown circumstances. It seems clear, then, that T strain Mycoplasma are one of the major causative agents in NGU. One authority has anticipated that future work will show 70% of venereally acquired non gonococcal urethritis to be due to T strains.

Attempts have been made to relate Mycoplasma to Reiter's Syndrome. There is no current evidence, however, to support the claim for a direct association. T strain Mycoplasma are isolated as frequently in Reiter's Syndrome as in normal controls, joint fluid cultures are negative, and the syndrome is unresponsive to tetracycline or erythromycin. However it is always possible to postulate an indirect relationship between Mycoplasma and Reiter's Syndrome, similar to that between group A streptococci and rheumatic fever, in which cultures from arthritic joints are negative for streptococci, and peni-

cillin has no effect on the established disease. Support for this line of reasoning comes from reports of temporal relationships between Reiter's Syndrome and Mycoplasma associated NGU.

In other types of genital infection the role of Mycoplasma is more widely accepted. Dienes and Godsall first isolated (in 1937) what was probably a *Mycoplasma hominis* from a Bartholin abscess. Other more recent reports have established an unquestionable etiologic role for *Mycoplasma hominis* in a formidable list of genital infections: acute salpingitis, pelvic peritonitis, infected Caesarian section wounds, inflamed prolapse, post natal pyrexia, and more importantly in puerperal sepsis and spontaneous abortion. *Mycoplasma fermentans* has been isolated from a few patients with penile lesions and with vulvovaginitis but the significance of these findings is unsettled. A recent study has shown T strains to be associated commonly with human abortions, earlier premature deliveries and increased fetal wastage.

DERMATOLOGICAL DISEASE

The main interest in the association of dermatological disease with Mycoplasma infection has centred upon the relationship between Mycoplasma and erythema multiforme – both the major (Stevens Johnson) and minor (Hebra's Disease) forms. Some of the features of erythema multiforme, such as the prodromal malaise and fever, the self-limited course, and the recurrences suggest that infection may play a part. Cases may follow clinical herpes simplex infection and smallpox vaccination. Orf, ornithosis, adenovirus and histoplasmosis are also known to precipitate erythema multiforme. Several studies have now shown Stevens-Johnson Syndrome to be associated with *Mycoplasma pneumoniae* infection. In febrile muco-cutaneous syndrome associated with acute respiratory disease, *Mycoplasma pneumoniae* is probably the single infective agent most commonly recovered. In fact up to 50% of these may be caused by *M. pneumoniae*. Despite this the frequency of Stevens Johnson Syndrome following *Mycoplasma pneumoniae* pneumonia is extremely low. In patients with erythema multiforme presenting to dermatologists, however, it is very unlikely that *Mycoplasma pneumoniae* plays such a prominent role.

Lyell has studied erythema multiforme minor in association with *Mycoplasma pneumoniae* infection. *Mycoplasma pneumoniae* was isolated from skin blisters of only a small minority of the more severe cases whose sera showed a rising titre of complement fixing antibody. The pathogenesis of the skin lesions in erythema multiforme is not clear. Demonstrating organisms in skin blisters suggests an actual skin infection rather than an allergic reaction to the infection. The latter is not entirely ruled out, however, because the eruption could be due to a hypersensitivity to *Mycoplasma pneumoniae* antigen in the skin.

Erythema nodosum, pyoderma, abscesses and pityriasis rosea are the only other dermatological lesions with proven association to *Mycoplasma pneumoniae*. Reports of this association are few and further surveys are required to more clearly define the role of Mycoplasma in these conditions.

Grace, has seen bodies that he interprets as Mycoplasma in pemphigus vulgaris and vegetans. He suggests that the Mycoplasma may act as antigen, stimulating production of skin auto-antibodies. Such antibodies are detectable in pemphigus. Cold agglutinins – found in both Mycoplasma infections and Stevens Johnson Syndrome (and in many viral diseases) – are auto-antibodies against red cell I agglutinogen – and thus if Mycoplasma induce production of these auto-antibodies, it is theoretically possible that Mycoplasma infection could result in the development of auto-antibodies to epidermis. Clearly many gaps remain in our knowledge of Mycoplasma related skin disease.

RHEUMATOID ARTHRITIS AND LUPUS ERYTHMATOSUS

The possible role of Mycoplasma in the pathogenesis of rheumatoid arthritis is another hotly contested issue. *Mycoplasma fermentans* has now been recovered with some regularity, from the joints of patients with sero-positive rheumatoid arthritis. No such recovery has been made from patients with sero-negative arthritis. Antibody to *Mycoplasma fermentans* is significantly more prevalent in the sera from patients with sero-positive rheumatoid arthritis than in sero-negative arthritis or in normal control groups. On the other hand normal serological tests for Mycoplasma antibodies such as haemagglutination inhibition have not clearly differentiated rheumatoid arthritic patients from controls. To overcome this, Williams and associates have used the leukocyte migration inhibition test. This is based on the finding that leukocytes from sensitized individuals do not migrate enthusiastically from a capillary tube in the presence of an antigen to which the individual is sensitive. It was found that leukocytes in 67% of patients with proven rheumatoid arthritis were inhibited by a preparation of *Mycoplasma fermentans* membrane. Inhibition of the migration of leukocytes from osteoarthritic patients or healthy control subjects was not observed.

There is still some uncertainty regarding the interpretation of the migration test, but it is believed to be a manifestation of cell-mediated (delayed-type) hypersensitivity. Furthermore, intradermal injection of Mycoplasma membranes evokes mixed Arthus and delayed type reactions in rheumatoid arthritics. These results suggest that rheumatoid patients uniquely exhibit hypersensitivity to the antigens of *Mycoplasma fermentans*. Intra-articular injection of antigen into animals leads to a clearcut arthritis with many features of the human condition. Thus it is postulated that *Mycoplasma*

fermentans combines in some fashion with host IgG. This altered protein then provokes the formation of auto-antibody (rheumatoid factor), which in turn causes the joint to be inflamed because of the active antigen-antibody reactions going on inside. It may be more than coincidence that of the many strains of Mycoplasma studied, only *Mycoplasma fermentans* has this ability to bind immunoglobulins so avidly. The periodic exacerbations in chronic rheumatoid arthritis could be blamed on *Mycoplasma fermentans* if at times the organism persisted within synovial cells (protected from immunological attack), and at other times was present extracellularly where it provoked the hypersensitivity response. Remissions could occur in individuals who developed humoral and/or cell mediated immunity to the organism. Further argument in favour of a link between *Mycoplasma fermentans* and rheumatoid arthritis is the fact that gold salts and antimalarial drugs which have a beneficial effect in many cases of rheumatoid arthritis strongly inhibit the growth of this organism.

In another experiment, Williams demonstrated the presence of Mycoplasma-like bodies by electron microscopy in 85% of synovial effusions from sero-positive rheumatoid arthritis. By means of a new culture technique *Mycoplasma fermentans* was isolated from 50% of these patients. Results were negative in sero-negative arthritics and in osteoarthritic patients.

Stevens has drawn attention to resemblances between the increased immunoglobulins in SLE and the serum changes in animals with chronic infections or in those hyperimmunized with bacterial nucleoproteins and polysaccharides. Stevens goes on to suggest that antibody (or immunologically competent cells) directed towards the polysaccharides of an infecting micro-organism crossreacts in Lupus with similar chemical groupings of the polysaccharides in body membranes. This is analogous to rheumatic fever and acute glomerulonephritis in which antibodies are formed against similar antigenic determinants present in cell walls of certain serotypes of *Strep. pyogenes* and also in heart muscle sarcolemma, synovial membrane and kidney basement membrane. Thus the immune reaction to a chronic infection rather than the chance emergence of "forbidden clones" of immunologically competent cells, active against the body's own tissues, is seen as the underlying pathological process. It is of some interest therefore, that Bartholomew has isolated Mycoplasma from the tissues of 4 of 6 patients with Systemic Lupus.

In studies with findings and interpretations contradictory to those of Williams, Sharp used synovial fluid from patients with Reiter's Syndrome and patients with rheumatoid arthritis. He failed to isolate Mycoplasma from these patients and failed to find rising antibody titres to Mycoplasma in these patients. He concludes that at the moment it would seem unwise to accept an etiologic role for Mycoplasma in the articular diseases of man. The matter is far from concluded and it seems certain that in the future much more interesting work will be done on the subject.

NEUROLOGICAL DISEASE

Landry-Guillain-Barré Syndrome (infectious polyneuritis) often has a preceding history of an infectious disease; a third of these infections are of the upper or lower respiratory tract. Several cases of infectious polyneuritis associated with a preceding *Mycoplasma pneumoniae* infection have now been reported. It is suspected by several authors that *Mycoplasma pneumoniae* infection can affect any level of the nervous system and in line with this, reports of meningo-encephalitis, perceptual deafness and acute cerebellar ataxia following *Mycoplasma pneumoniae* infection have already been published.

LEUKEMIA AND NEOPLASMS

The isolation of Mycoplasma from some cases of leukemia, lymphoma and from certain solid tumours, although of uncertain etiological significance is the subject of intense interest at present. Workers have occasionally found virus-like particles in electron microscopic studies of the lymph nodes, bone marrow or plasma of human patients with leukemia or lymphoma. Some of these particles have a double plasma membrane and their cytopathic effect (CPE) in tissue culture is inhibited by tetracycline. Hummeler suggests that these particles are different sized elements of Mycoplasma. In 1963, Murphy isolated these organisms and found them to be a Mycoplasma. Negroni (1964) confirmed this work as have many subsequent researchers.

All the reports have very carefully avoided attributing an etiological role to the Mycoplasma apparently isolated. The disturbed defense mechanisms of the leukemic patient are quite familiar and the obvious explanation is that Mycoplasma colonize the bone marrow and other organs as a result rather than as a cause of the disease. It would be premature however, to conclude that the obvious explanation is necessarily the correct one and more evidence is required. Several workers have found that Mycoplasma infections in some cells reduced chromosome numbers and increased chromosome aberrations. These may be caused by the effect Mycoplasma have on pyrimidine synthesis, or by the action of Mycoplasma nuclease on cell DNA. Thus a possible mechanism for Mycoplasma induced neoplastic change exists.

CONTROL OF MYCOPLASMA INFECTIONS

Once in vitro culture techniques were well established for Mycoplasma it was possible to determine precisely the spectrum of sensitivity to a variety of antibiotics. The organism is exquisitely sensitive to erythromycin and to most of tetracycline drugs. (*M. hominis,* however, is resistant to erythromycin. In this respect it is different from other Mycoplasma species.) Studies using

these drugs correlate well with clinical findings (after 7 days of treatment) of decreased number of days with fever and a decrease in the time required for radiological improvement.

The present practice is to treat *Mycoplasma pneumoniae* pneumonia for 7 days. However, incomplete relief of cough, occasional relapse after short courses of treatment and continued shedding of the organism often characterize this type of treatment. Two to three weeks of treatment might alleviate some of the problems encountered with seven days of treatment. Because many of the varied agents responsible for non-bacterial pneumonia, in addition to M. pneumoniae, respond to tetracycline this drug provides good coverage in situations where the precise aetiology cannot be ascertained clinically or bacteriologically. This is because tetracycline is also effective against pneumonia caused by psittacosis and Q fever, which cannot be differentiated from the others on clinical grounds. One gram daily should be given for two to three weeks. In pediatric and pregnant patients, obviously erythromycin would have to be the drug of choice.

Mycoplasma pneumoniae has variable sensitivity to kanamycin but is quite resistant to penicillin as would be expected with the absence of a cell wall. These findings have been confirmed in studies of NGU. Animal experiments with *Mycoplasma pneumoniae* have shown these antibiotics to be effective when administered 15-20 days after inoculation; but when administered soon after inoculation the rate of occurrence of pneumonia was not significantly altered. This suggests that prophylatic treatment of *Mycoplasma pneumoniae* contacts would not be effective. Human experiments have so far provided equivocal results.

Pneumonia patients infected with *Mycoplasma pneumoniae* appear to develop both IgM and IgG antibodies. The antibody persists in readily measurable quantities for 6-12 months, but falls off thereafter. Patients infected with *Mycoplasma pneumoniae* show two particularly interesting antibody responses: cold agglutinins and Streptoccocus MG agglutinins. It seems that Streptococcus MG agglutinins are the result of a fortuitous possession of a cross reacting antigen between the two organisms. Cold agglutinins have been discussed previously.

Although antibiotics are effective in reducing morbidity due to *Mycoplasma pneumoniae* infections they are not the answer to community control of these infections. Both inactivated and live attenuated vaccines have been worked on. Inactivated vaccine induces the development of growth inhibiting antibodies in only about half of those vaccinated. In some studies, post-vaccination inoculation of *Mycoplasma pneumoniae* failed to protect many of the vaccines, while others who failed to respond to the vaccine (by producing antibodies) developed severe clinical illness. These findings suggest that those who failed to develop antibodies may have been sensitized and had their disease made worse. The production of a live attenuated vaccine for humans is presently under way. Live attenuated vaccine has been used however, with much success in animals. Also being developed is a live virulent, but cold mutant vaccine. Given intranasally this mutant strain can multiply at 35°C in the nasal epithelium but cannot at 37°C in the tracheal or pulmonary tissue. This produces good antibodies without producing clinical disease.

CONCLUSION

In conclusion, although the role of Mycoplasma in causing pneumonia seems well established, its place in the etiology of many other conditions is far from secure. It seems certain that Mycoplasma will come to be recognized as a prime cause of non gonococcal urethritis and probably of many abortions, and that it will be implicated in the pathogenesis of Stevens Johnson Syndrome and infectious polyneuritis. Whether it has any role in the etiology of autoimmune collagen disease or leukemia remains an open question.

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Idiopathic Scoliosis Of Childhood

Barry W. Malcolm 7T1

In the paediatric age group, no orthopaedic deformity is as complex and frustrating as scoliosis – lateral curvature of the spine. Although the word scoliosis was first used by Hippocrates and his contemporaries, it was Paré in the sixteenth century who equated scoliosis and lateral curvature of the spine. Other writers of the past branded the kyphoscoliotic or 'hunchback' as sinister and macabre, and it was Victor Hugo who immortalized this deformity in his novel, "The Hunchback of Notre Dame."

The past sixty years have witnessed the greatest advancements in etiology, pathophysiology, diagnosis, prognosis and treatment, which in turn have been dependent on advancements in genetics, anesthesia, hematology, radiology, pulmonary function, mechanics, and operative technique. In this review, I would like to discuss scoliosis, with emphasis on the most common variety, idiopathic scoliosis of childhood.

The typical deformity of scoliosis is very rarely a pure lateral curve, but usually contains a rotational element which may be superimposed on a normal or abnormal kyphus or lordotic curve.

CLASSIFICATION:[1,2]

a) Nonstructural:
i.e., a curve totally correctable by forward bending, recumbency, or traction; with no fixed rotation of vertebral bodies or spinous processes.
   i) postural
   ii) compensatory
   iii) sciatic
   iv) inflammatory
   v) hysterical

b) Structural: i.e., a curve which has irreversible components
   i) idiopathic (85%)
   ii) neuropathic – congenital
      – meningomyelocele
      – Friedreich's Ataxia
   – acquired
      – cerebral palsy
      – syringomyelia
      – poliomyelitis
      – traumatic paraplegia
   iii) osteopathic – congenital
      – vertebral anomalies – eg. hemivertebra
      – osteogenesis imperfecta
      – Marfan's syndrome
   acquired
      – osteoporosis
      – adolescent kyphosis
   iv) myopathic – congenital
      – muscular dystrophy
      – arthrogryposis
      – amyotonia congenita
   v) thoracogenic – fibrothorax
   vi) metabolic – rickets
   vii) radiation
   viii) infective – tuberculosis of the spine (Pott's disease).

TERMINOLOGY

Presently, the Scoliosis Research Society of North America is revising much of the terminology and nosology used in the scoliosis literature. For example, the terms primary and secondary curve, which were at best difficult to define (especially in an individual with two curves), have been discarded in favour of the terms major and minor curves. The curves of greatest importance, insofar as magnitude and structural elements are concerned, are called major curves, the others minor curves. An individual's spine may have two or more major curves.

a) Compensation

Scoliosis was said to be compensated if a plumb line from the symphysis menti passed through the symphysis pubis. This term has been qualified clinically and radiographically, such that, in a compensated curve, the shoulders are level and the lateral costal margins are parallel to the iliac crests in an anteroposterior radiograph of the spine. Compensation results from formation of minor curves above and below the major curve or curves.

b) Curve Type

Scolioses may be characterized by their shape, location, direction of deviation and age of onset. The accurate assessment of any curve by these criteria, is important for both prognosis and treatment.

   i) shape – 'S', 'C', double 'S'
   ii) location
      – cervical
      – thoracic
      – thoraco-lumbar
      – lumbar
      – combinations
   iii) direction of deviation – convex to the right or to left, depending on the direction of the major curve
   iv) age of onset –
      infantile: from birth to three years of age
      juvenile: four years to end of the ninth year
      adolescent: ten years to the end of skeletal growth

c) Angle of Curvature

Perhaps the most crucial aspect of the diagnosis, prognosis and subsequent treatment of scoliosis, is the accurate measurement of the curve at the initial, and following examinations. The accepted method is that described by Cobb in 1948.[4] This consists of selecting and labelling by number the end vertebra of each curve,

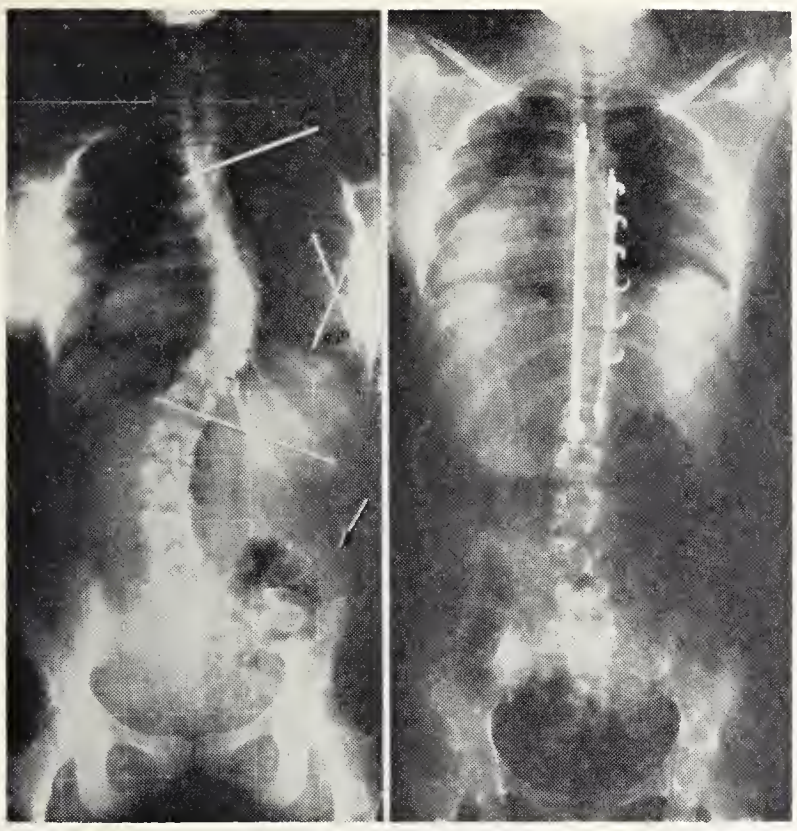

FIGURE 1: *The preoperative radiograph on the left shows a 45° right thoracic scoliosis, as measured by the Cobb method. The arrow points to Risser's apophysis, which has not fused to the ileum. The postoperative film on the right shows a posterior spinal fusion, with the Harrington distraction rod and compression apparatus in place producing an excellent correction.*

where these vertebrae are maximally tilted. (Rotation is important and can be measured, but is not utilized in this particular system.) Lines are drawn from the inferior margin of the caudad vertebra, and from the superior margin of the cephalad vertebra, such that the greatest magnitude of the curve is produced. Since the point of intersection is in the concavity of the curve, usually off the radiograph, the angle of intersection of perpendiculars from the superior and inferior lines equals the angle of curvature in degrees. (see figure 1)

It is absolutely imperative, that any further radiographic examination of the spine be compared to the original radiograph, and that the identified vertebrae be used consistently in curve measurement.

d) RISSER'S APOPHYSIS

This iliac apophysis begins its development on the antero-lateral aspect of the ilium, as seen in the A-P radiograph. By the time this elongated ossification centre has grown posteriorly and medially to the sacro-iliac joint and has fused to the ilium, vertebral growth is virtually complete. This relation was first noted by Risser in 1936, and bears his name.[5] The average age at completion is 14 years in the female and 15½ years in the male. The importance of this physiologic sign lies in its ability to predict the probable duration of spinal apophyseal growth and therefore progression of spinal curvature.

Spinal growth and curve progression may continue for eighteen months to two years after closure of the iliac apophysis. This growth may be gauged by the ring apophyses of the vertebral bodies and the proximal vertebral epiphyses of the thoracic ribs.

Since scoliosis progresses with growth, a sign such as this on routine radiographs is very useful. (see figure 1)

INCIDENCE AND ETIOLOGY

Scoliosis of some degree is present in one out of twenty people. Of this number, idiopathic scoliosis accounts for more than 85%. Scoliosis and related problems are responsible for 50% of all orthopaedic admissions to the Hospital for Sick Children, Toronto.

Girls are more commonly affected in the juvenile and adolescent age groups; whereas, boys are involved more frequently in the infantile group. It is of interest that there is a higher incidence of the infantile variety in Great Britain and Europe, as compared to Canada and the U.S.A. The reason why the right thoracic curve is the most common adolescent curve is unexplained.

It has been known for some time that genetic factors are important. A study by Wynne-Davis (1967) in Edinburgh[6], revealed that the incidence of scoliosis in the families of girls with adolescent onset deformity, was almost twenty times greater than the general population. Similar more recent studies have supported the familial incidence; however, a specific pattern of inheritance remains to be elucidated. It presently appears to be a multigenetic inheritance with varying penetrance and expressivity.

Dr. P. Conen at the Hospital for Sick Children, Toronto, is presently examining cells and cell cultures taken from connective tissue biopsy at scoliosis corrective surgery. After thirty to forty days of growth, these cells possess altered shape and metachromasia when stained with a Blue dye. These cells are now being compared with normal cells and cells with proven chromosomal abnormalities, to determine the significance of these changes.[7]

In spite of investigation through research, the precise etiology of idiopathic scoliosis remains an enigma.

PATHOPHYSIOLOGY

The majority of idiopathic scolioses begin as small flexible curves in previously straight spines; although, a few of the progressive infantile variety have shown structural manifestations at birth.

The commonest curve occurs in the juvenile and adolescent age group. It is most frequent in girls, convex to the right, and has end vertebrae at T_2 - T_4 to T_{11} - T_{12}. It would appear then, that this would be the ideal curve to describe. As the curve progresses, there is, at first, a loss of flexibility, which may be observed

on side bending examination and by radiographs. There are no asymmetrical changes in the thoracic cage at this early stage. Soft tissue contracture develops, and as growth continues and the curve increases, unequal pressure is exerted on the vertebral ring apophyses and cartilagenous growth plates. This may lead to inequality of growth rate on the convex and concave sides, ultimately producing vertebral wedging (Heuter-Volkman Law of partial epiphyseal plate pressure). At this point, the curve becomes structurally irreversible. The compensatory curves which develop around the major curve, may progress to vertebral wedging and irreversibility. A recent study has concluded that some deformities are not due to asymmetrical endochondral growth, and postulated that some extraosseus cause – biological, physical, or both – may be responsible. The etiology remains obscure!

Vertebral rotation occurs in the major curve, such that the spinous processes are rotated toward the concavity of the curve. The ribs on the convex side are carried posteriorly with rotation, producing the classical hump seen on forward bending; and the ribs of the concavity are pushed anteriorly causing another variety of presenting signs, such as a prominent breast with anterior chest deformity.

The structural changes in the thoracic cage produce deleterious effects on the cardiorespiratory system. The decreased thoracic mobility may produce a restrictive hypoventilatory abnormality. The pulmonary function studies reveal a significant decrease in vital capacity and maximum breathing capacity, when compared to normal values. Some studies have suggested a change in compliance of the chest as a cause for these results, but, there is not complete agreement on this topic. Chronic alveolar hypoventilation may lead to pulmonary hypertension and kyphoscoliotic heart disease with eventual cor pulmonale. Not all patients with scoliosis develop these severe changes. Surgical correction of the deformity does not always produce major measurable changes in the restrictive abnormality. In the more severe degrees of scoliosis and respiratory inadequacy, there is often a dramatic pulmonary improvement with increased exercise tolerance and diaphragmatic excursion. The long term effects of such correction must wait further study.

Radicular and peripheral nerve involvement is virtually unknown in idiopathic scoliosis. Pain too, is not a problem, until the late 40's-50's, when, in untreated cases, painful degenerative joint disease of the zygoapophyseal joints supervenes.

Pathological changes in the spinal cord, nerve roots, and microcirculation remain to be explored.

Slow progression of curves after cessation of spinal growth is thought to be due to wedging of the intervertebral disc spaces. This change may amount to one degree per year; however, this does not appear to explain the problem of adult scoliosis.

DIAGNOSIS AND PRETREATMENT ASSESSMENT

On the basis of clinical awareness and a complete history and physical examination, the family physician can usually make a presumptive diagnosis in early scoliosis. On the average, a curve must be greater than 25 degrees before it is clinically evident. After diagnosis, the patient should be referred to an orthopaedic centre, where further radiographic and ancillary examinations may be undertaken, and treatment prescribed. There is no place for procrastination or a "maybe you will grow out of it" attitude!

a) clinical suspicion
   positive family history
   shoulder asymmetry
   prominent scapula, breast, or hip
   rib hump posteriorly
   curve, if greater than 25 deg.

b) physical examination – important to exclude known systemic diseases and malformations which may produce a curve
   complete neurological examination
   leg length mensuration for comparison
   musculo-skeletal exam – Right and Left lateral bending of the spine to determine flexibility, and forward bending to determine rotation.
   gait
   café au lait spots

c) radiographic examination – 3′ standing and supine A-P radiographs of the spine – always compare with initial radiographs, use the same vertebrae to measure the angle of curvature

d) other –
   pulmonary function tests

TREATMENT

Aims – prevent progression
– correct deformity
– stabilize deformity

Scoliosis treatment may be operative or non-operative; at the same time it may be radical or conservative. For example, a six-year-old girl could wear a scoliosis brace for the duration of her skeletal growth, i.e. 10-11 years. This treatment is considered radical because of the restriction of normal activities and the problems inherent in complex bracing. In comparison, surgical treatment may allow activity after a period of 6 months.

a) Non-operative

i) *Continuous followup, with no specific treatment:*

This may be deemed the treatment of choice, depending on the curve type, age of the patient, and the age at onset. During the entire period of skeletal growth

however, these patients should have radiographic examination of the spine at three monthly intervals. After cessation of growth, the spine should be examined biannually, progressing to annually. This is the desirable routine in all post-operative patients.

ii) *Recumbency and exercises:*

This mode of treatment was used extensively in Europe until five or ten years ago. The American Orthopaedic Association, in 1943[3], reviewed several thousand cases of scoliosis treated by exercises. They concluded that active exercises were ineffective in controlling curve deterioration or improving the existing curve, when assessed by measurement of the curve. This statement is endorsed by orthopaedic surgeons in Toronto.

iii) *Milwaukee Brace:*

This brace, first used by Blount in 1944, and described in the literature in 1958[8], remains the most effective bracing technique for treatment of scoliosis. (see figure 2)

It provides distraction of the spine by means of a moulded leather pelvic girdle, and a mandibulo-occipital assembly, with vertical bars between. Pressure necrosis of the skin is not a problem of the pelvic girdle, because of its large skin contact; however, after several years in the brace, the head apparatus may produce pressure atrophy of the soft tissues of the mandible with protrusion of the upper incisors, which may or may not resolve spontaneously after removal of the brace. Recently, it has been recognized that the closer fit of the occipital rest is of greater value in distraction than the chin pad; and more recently patients are being fitted with a moulded throat pad which does not produce mandibular deformity.

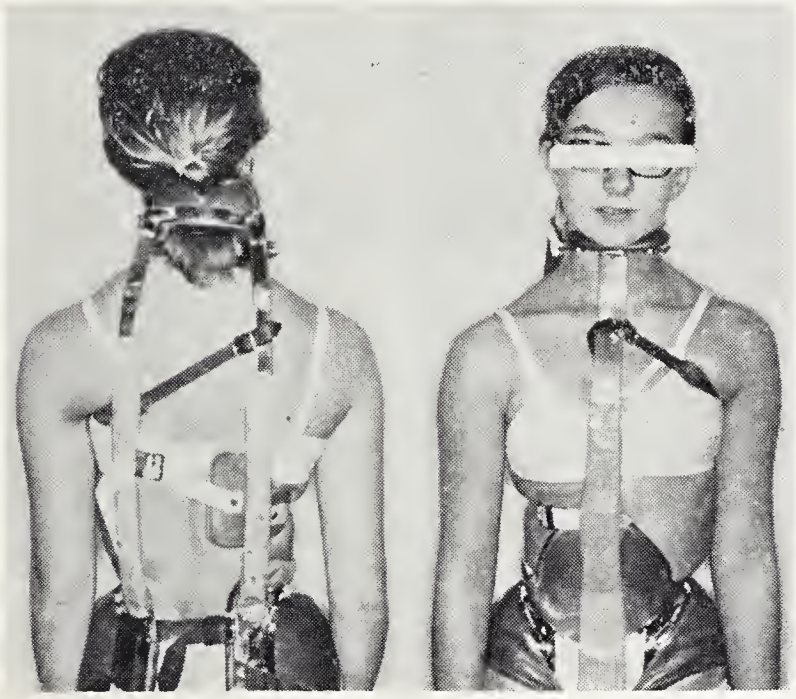

FIGURE 2: *Shows anterior and posterior views of a Milwaukee Brace.*

Lateral pressure is provided by adjustable pads attached to the vertebral bars. The pad is placed over the lateral ribs leading to the apex of the curve, and pressure is then exerted via these ribs on the spine. This apparatus does not lend itself to use on upper thoracic, cervical, or lumbar curves.

The brace is worn continuously until spinal apophyseal growth is complete, being removed only for bathing. This rule may vary with age of onset, size of curve, cosmetic deformity and the inclination of the surgeon. The braces are adjustable for growth, and must be refitted every two or three years.

Indications:
- — control of patients too young for surgery until they are suitable for surgery
- — control of scoliosis in adolescents who develop their curves near the end of the growth period.

Clinical evaluation has consistently supported the use of the Milwaukee Brace in the treatment of idiopathic scoliosis.

iv) *Serial casts:*

Serial casts may be used to prevent progression of the curve in the very young child who has progressive infantile scoliosis. They are frequently able to ease out of a brace, by constant movement. This treatment is time consuming, and involves prolonged confinement, often in bed, precluding a growing child's normal activities.

b) OPERATIVE

The aims of operative treatment can only be achieved by fusion, which alone may prevent progression and stabilize the spine, and instrumentation, which offers excellent reduction in the angle of curvature. The treatment may be fusion alone, or, instrumentation coupled with fusion.

Rotation is the most obvious and most difficult deformity to correct. Many operations were devised to reduce the cosmetic deformity of the rib hump associated with rotation, including resection of the involved ribs; however, these have proved to be futile.

The success of spinal surgery is limited by a number of factors. These include:

- — the age of the patient and therefore the developmental stage of his vertebral bodies and posterior elements. Under the age of five years, the latter are cartilagenous, thus precluding certain types of instrumentation.
- — the presence or absence of vertebral bodies or posterior elements eg. meningomyelocele

— general health of the patient
— the experience and proficiency of the surgeon.

i) *Fusion-in-situ:*

In children less than six years of age, with rapidly progressive curves, who are unsuitable for bracing and too young for instrumentation because of vertebral immaturity, a fusion-in-situ may be undertaken, to prevent progression of the curve. It is then possible, in a few years, to revise the fusion by osteotomy of the graft and instrumentation to produce correction, and fusion to maintain it.

ii) *Cast correction and fusion:*

Two techniques by plaster correction are presently employed in some centres. Both were described by Risser.

a) The Risser hinging jacket[3] is a spinal plaster, applied to the patient while on a traction frame. When dry, a hinge is placed on and incorporated into the plaster on the convexity of the curve, and a turnbuckle is applied in like manner on the concave side of the curve. The plaster is divided circumferentially at the apex of the major curve, and over the next few weeks correction is produced by turning the turnbuckle. There is slow but efficient correction. To obtain maximal stress on the lower segment, the lower limb on the convex side is placed in the spinal jacket.

b) The Risser localizer jacket[3], is applied to the patient on a specially designed scoliosis table, much like a medieval torture rack. It comprises a cervical portion, well moulded about the mandible and occiput, and a pelvic section, moulded around the iliac crests. By means of a halter around the neck and traction bandages attached to the pelvic cast, the child is stretched to his limit of tolerance. When correction is maximal, the superior and inferior casts are joined by more plaster bandages to make a body cast. Lateral pressure is applied to the ribs on the apex of the curve by means of a localizer until the cast is dry.

Operation, after correction in these casts, is carried out through a large posterior window. This aperture exposes the entire spine and both iliac crests – the donor site for cortical bone grafts.

Although effective, these procedures are very cumbersome. Post-operative immobilization in the cast varies from three to six months. Better padding under casts has reduced the incidence of pressure necrosis of the skin, and various anterior windows have made anesthetic intubation easier and permitted easier access to the precordium in case of cardiac arrest.

iii) *Halo-femoral traction:*

Halo-femoral traction is applied to a patient on a Stryker frame. Using either a skull tong, or more recently a stainless steel hoop, attached by screws into the supra-orbital ridge and occiput, distraction weight of up to 30 lbs. is applied to the head for 2-3 weeks. Kirschner-like wires through both distal femora, attached to a similar weight, complete the traction apparatus.

The use of these preoperative traction methods will usually attenuate soft tissue contractures, and markedly reduce a curve in about two weeks, at which time instrumentation and fusion may be undertaken with greater ease and safety.

iv) *Instrumentation and Fusion:*

In 1949, Harrington[9] began experimenting with internal fixation of the spine. He published his first results in 1966.

The Harrington procedure requires extensive exposure of the vertebral column, including portions above and below the major curve. The paravertebral muscles are elevated and retracted by subperiosteal dissection from the spinous processes and laminae, to the tips of the transverse processes. To provide the distraction force, two hooks and a long rod are used. One hook is placed around the upper lamina of a vertebra above the major curve on the concave side; another on the concave side of a vertebra below the major curve. Using a special tool, the two hooks are distracted by a rachet action and maintained by the rod. The distraction pressure may reach 300 p.s.i. A compression rod, with hooks on each transverse process, is applied to the convex side of major curve vertebra, and tightened. With maximum correction obtained, spinal fusion using iliac bone grafts is carried out.

The patient is nursed on a Stryker frame for 2 weeks, in a body jacket for 3 months, in bed, and then ambulatory in a light removable plastic brace for another three months. There is a gradual resumption of activity as tolerated with virtually no limitations at the end of one year. The rods remain in situ forever.

The results of this operation are most encouraging, and it remains the procedure of choice in Toronto.

Complications of this major surgery are few. Pseudarthroses of the fusion do occur, but the incidence has been reduced by meticulous surgical techniques so that in idiopathic scoliosis surgery it is virtually unknown even with such a long spinal fusion.

FUTURE

The enigma of scoliosis requires a great deal of research and experimentation. The pathophysiology on a miniscule level is grossly unknown. The contributions of biochemistry and genetics have yet to be adequately assessed.

In the past couple of years, there have been great advancements in the surgical treatment of scoliosis. Since the indications for these procedures are still being assessed, I will only briefly mention them.

The halo-pelvic distraction apparatus employs a stainless steel hoop attached to the skull by screws, and another hoop encircling the pelvis. It is attached by

threaded screws through the iliac wings. Adjustable distraction rods, are attached to the hoops, and correction can be produced and maintained. It is virtually pain free, and permits almost completely normal activity while in traction.

The Dwyer procedure 1969[10], involves anterior exposure of the vertebral bodies by a large thoracoabdominal incision, splitting of the diaphragm, and retraction of abdominal viscera and major vessels. The vertebrae to be fused, are indentified. A metal plate, and large bore screws are attached to the convex side of the vertebral bodies of the major curve. Then, using a flexible cable firmly attached to an end-screw and passing through the heads of the other screws, a Tensioner device approximates the screw heads, and a swaging clamp fixes the head firmly to the cable to maintain the force. As this process progresses step by step down the length of the curve, it is gradually straightened. Removal of the intervertebral discs is necessary to achieve maximal correction and anterior interbody spinal fusion.

Postoperative management of these patients is presently similar to that of the Harrington instrumentation.

The anterior spinal osteotomy, which may end up being a total circumferential osteotomy of the spine, is reserved for extremely severe localized curves. It has no standard technique and is designed for each specific patient. These most complex operations have not yet had their full indications worked out.

The future holds many challenges in the total medical and surgical management of this fascinating problem.

CONCLUSION

Idiopathic scoliosis represents a very common orthopaedic spinal deformity. It can progress relentlessly with skeletal growth. With the use of modern diagnostic procedures and operative technique, the ultimate prognosis is gratifying.

ACKNOWLEDGEMENT

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While on the Pediatric rotation of the clinical clerkship at the Hospital for Sick Children, final year medical students in Toronto are assigned a Social Pediatrics project, usually in groups of three students. Ten different projects are available. Towards the end of the rotation, one member of each team presents a report on the project of the group. On Wednesday, March 3, 1971, W. Skala, S. Silverberg and the author visited the Ontario Mental Retardation Centre. The following is a personal account of the afternoon.

No. 2 Surrey Place is an old sturdy six-storey building sandwiched between Women's College Hospital, The Banting and Best Research Institute buildings and some tall brand new Government of Ontario buildings. We arrived at about 2.30 p.m. and were directed upstairs by a receptionist who told us that Dr. Bonkalo was expecting us. Dr. Bonkalo is an Associate Professor in Psychiatry in the medical faculty in Toronto and chief consultant in medicine at the Mental Retardation Centre. After a remarkably friendly welcome and introduction, the informal discussion began.

You graduate in three months, so I presume you have been thinking of what you will do. What are your career plans?

The answers came with some hesitation. One of the students wanted to be a good all-round doctor, another was going into the specialty of Obstetrics-Gynaecology, the third was vague enough to create the impression that he would specialize in something but is currently generally interested in medical and paramedical problems. Nobody said he would be a psychiatrist or paediatrician. The point was not belaboured. Even at this stage of our career we know that mental retardation is uncommon enough that no one is likely to see a new case everyday except the people who work in special centres dealing with the problem. Yet it is common enough that we can be confronted with it on any day in the practice of the art and science of medicine – from the family physician who hears the perturbed mother complain that the child is slow to the clinical biochemist who detects phenylketonuria in the laboratory. What makes the problem even more worrisome is that mental retardation is not always hereditary. Even a brilliant doctor can have a mentally defective child in his family. If this happens the child has a greater than normal chance of being severely defective as will be explained later.

The next question came. How much did we know about the problem of mental retardation? It was a difficult question to answer and the replies were necessarily vague. However, at the end of the day the answer was obvious . . . very little. Our understanding of the medical aspects prior to our visit was probably adequate but there is more to mental retardation than meets the eye in reading a standard Paediatrics text.

From our informal discussion with Dr. Bonkalo, we had some major points re-iterated.

The definition of mental retardation entails three major facets. The first is that the mentally retarded has an intellectual capacity below the accepted limits of normalcy for his group. Secondly, there must be an element of arrest of intellectual maturation (i.e., the appropriate I.Q. for age was never reached). This contrasts with dementia where there is intellectual deterioration after a full I.Q. has been reached. Examples of the latter are well known clinically. One such syndrome is Huntington's Chorea, an autosomal dominant trait which has as one of its features a deterioration in mental ability in the second and third decades of life, the individual having previously attained full intellectual maturity. The third point of note is that mental retardation may have known organic or idiopathic causes. The patient's age is irrelevant. Provided that the first two criteria are met, the diagnosis can be made whether the patient is two or twenty-two years old.

When the Intelligence Quotient, (I.Q.), of any culturally distinct group is plotted against the frequency of occurrence of the I.Q., an approximately Bell-shaped normal distribution is obtained. The mean for the population is arbitrarily fixed at 100. One standard deviation is $\pm$ 15. Naturally in considering mental retardation the problem lies with people on the negative side of the mean. Again arbitrarily, people within one standard deviation on the negative side of the mean are considered normal. Thus, the problem of retardation arises when the individual's I.Q. is 85 or less.

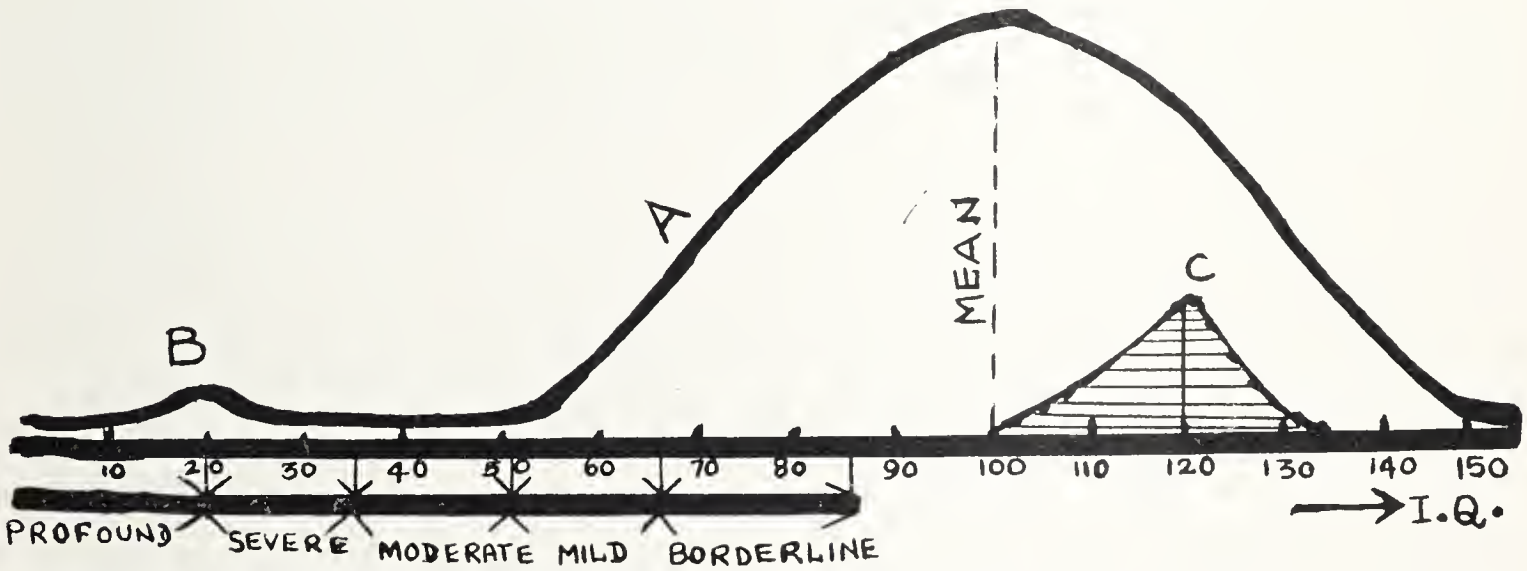
Various names have previously been assigned to the different degrees of retardation. In 1958, at an International Conference, a new classification was proposed, based on the I.Q. This is presented in the table below. The I.Q. is defined as the measured mental capability as compared with the mean ability of people of the same age with the individual.

Classification of Mental Retardation

I.Q.	Internatoinal Classification	Equivalent	Old Term
68-85	Borderline		
52-67	Mild	Mental Age	Feeblemindedness; moron
36-51	Moderate	8-12 years	Imbeciles
20-35	Severe	0-2 years	Imbeciles
<20	Profound	0-2 years	Idiots

This table is also expressed graphically below to illustrate the approximate relative frequencies.

MENTAL RETARDATION & THE GENERAL POPULATION



Since the mode of inheritance of intelligence is polygenic, the distribution shown above has some interesting features. In a group of individuals whose I.Q.s were recorded as 120, for example, the I.Q.s of their parents will be expected to cover a normal distribution designated as C in the diagram. Actual determinations have borne this out. Similarly, the I.Q.s of the parents of people declared retarded are found to fit into a normal distribution representative of their cultural community. The message is that the majority of mentally retarded children are born of clearly normal couples. The aetiology of retardation in such children is often genetic.

The little peak shown as B on the general population distribution represents the severely retarded individuals whose problems are caused by accidents – birth trauma, head injury, acute severe intrauterine and extrauterine infections or accidents, and the like.

The aetiology of retardation in this group has no genetic components. This is the "idiot" type of individuals found in highly intelligent people – highly skilled professionals, executives – like the readers of this article.

Next, we discussed the aetiology of mental retardation. The time-tested Congenital-Acquired classification would suffice, but Dr. Bonkalo suggested a different version, based on a chronological order. The list is long and unnecessary for this presentation. The scheme took the following format.

The earliest stage in the chronological order is pre-fertilization. These are genetic conditions predetermined in one or both parents – familial idiocy (Tay-Sach's disease), phenylketonuria and the whole realm or inborn errors of metabolism, only a fraction of which have mental retardation as a feature.

In the early post fertilization stage, abnormalities of chromosomal behaviour are prominent. Examples of these are Mongolism due to nondisjunction (trisomy 47), and translocation (another form of Down's syndrome). At later stages of embryonic life intrauterine infections, particularly viral (e.g. rubella), chemical and drug induced toxic states, organ hypoplasia or agenesis (thyroid), physical agents (radiation), are only some of the factors considered.

Difficult labour and puerperium, obstetrical accidents and head injury at birth are well known perinatal factors that can cause mental retardation.

In the immediate neonatal period, hyperbilirubinaemia has diverse origins, but has only one really dreaded consequence, that of *kernicterus*. At later stages of extrauterine growth and before full mental maturation is attained, head trauma and any of the encephalitides can intervene to produce an arrest of mental development.

Why do you suppose mental deficiency has lately assumed major importance in the field of mental health?

We supplied one answer to Dr. Bonkalo's question. It was correct, but not the only answer.

The advent of antibiotics and the overall generally higher quality of medical care has resulted in prolonging the life and reproductivity of people who are either overtly mentally deficient or have the potential of being so. Since some forms of intellectual impairment are hereditary, more mentally retarded people are born today. This is one factor. Increased emphasis on formal education within the last three decades has meant that a higher proportion of children now go to school or take instructions in highly technical jobs. Many borderline or mildly retarded people have a higher chance of being recognized today than in the days of their forefathers. They would function adequately as manual labourers, farmers, shepherds or lumber-jacks, but cannot learn Thermodynamics or Boelian Algebra. Many such borderline cases are usually recognised for the first time when they go to school or a vocational training centre. This is the other reason.

How does the Mental Retardation Centre get its clients and what can the centre do for them?

When mental retardation is diagnosed or suspected in an individual, ANYONE who knows about No. 2 Surrey Place can send the individual for assessment. Thus children and teenagers come to the centre usually on the initiative of somebody else – family doctors, paediatricians, psychiatrists, schools, vocational and social agencies, anxious parents or on the advice of friends and relatives.

The services available for the mentally retarded seem comprehensive: assessment, treatment, counselling (especially for the parents), and placement to appropriate environments. This could be a home, an institution or special agencies.

To provide these services, we were informed, the centre uses a multidisciplinary approach. On the regular staff there are psychiatrists, paediatricians, social workers, psychologists, geneticists, public health nurses and teachers specially trained to teach the mentally retarded.

What specific preventive measures can be taken to deal with mental deficiency?

This is a field with a lot of scope today. Definitive preventitive measures are few at present. Obviously the diverse nature of the aetiology implies that there is a lot of scope for research, ideally with multidisciplinary approach. Genetic counselling is a must, when appropriate. For example, familial idiocy (Tay-Sach's disease), is transmitted by a single recessive gene. Thus both parents of an affected child could be heterozygous for the condition though they are phenotypically normal. The chances of a child born to that couple manifesting the disease are only 1 in 4. Conversely, the chances of a child born to these parents being normal in genotype and phenotype is only 1 in 4. The conclusions are obvious. Niemann-Pick' disease (another one of the sphingolipoidosis), phenylketonuria and galactosemia all cause mental deficiency. They are all inherited as autosomal recessives. An important but rare cause of retardation is Tuberous Sclerosis which is inherited by an autosomal dominant gene with reduced penetrance. Manifested by epilepsy, mental defect, and a butterly-shaped rash of cutaneous nodules, fifty percent of the children of an affected individual will inherit the trait.

Apart from genetic counselling for the parents, other preventive measures are available. In the type of Mongolism caused by chromosomal nondisjunction or Trisomy 21, the child has 47 chromosomes. The frequency increases with maternal age at the time of the child's birth as shown in the table below.

Maternal Age	*Risk of Mongolism*
35 - 39	1:280 live births
40 - 44	1:70
45 - 49	1:40

If a woman in the high risk age group is pregnant and concerned about having a Mongoloid baby, it is justifiable and probably advisable to do amniocentesis and chromosome counts. A therapeutic abortion can be performed if there are 47 chromosomes and the woman wants an abortion.

Unfortunately, the cause of mental retardation in the majority of cases is simply not known. In these cases, prevention is impossible

It was now just past 4 o'clock. Dr. Bonkalo apologised for having taken so much of our time to complete his discussion then arranged for a tour of the building to be conducted by one of the social workers on staff.

Miss L. Marguiles, the social worker bubbled with enthusiasm as she conducted us around and told us about her job.

On the third floor, there is a recreation room where about a dozen children were watching *The Flintstones* on Channel 7. Two middle-aged women charges were in attendance, presumably to help keep order. The children ranged in age from about four years to twelve. Except for three with distinctly obvious but minor craniostosis, these children looked absolutely normal. Some were literally beautiful, lovable children. Many paid no particular attention to television but were engaged in individual activities. Some sat as if there were nobody and no objects around them. A few got curious and excited as our group entered the room, two dressed in the three piece hospital uniform, one in civilian attire, the fourth a social worker whom the children probably recognised.

An 8-year-old girl turned around and stood up to confront us.

Are you a doctor?

Yes.

She giggled. Two others turned around but did not get up. They asked that same question one after the other and got the same answer. The first girl seemed to want to know more.

Why are you here?

We would like to talk to you

Another giggle was followed by silence.

Our guide had other places of interest in the building to show us.

The "medical ward" is on the second floor. The first room had about 16 children's beds – a familiar sight on a standard paediatrics ward. All the beds were empty. As we entered the second section, we saw a two-year-old toddler who had trapped his head in a six-inch space under a four-foot door in an attempt to crawl out. Eight children ranging in age from about 2 to 8 years were playing with brightly coloured toys and stuffed animals, each by himself. Three of them had distinctly mongoloid features. I silently wondered why they were institutionalized. Dr. Bonkalo had said earlier that mongolism is in itself not an indication for putting the child in an institution. There must be some clear-cut medical indication. Perhaps the children also had congenital heart disease, leukemia, or were simply there for assessment, or to give the parents temporary relief from their domestic problems. Perhaps the child was there for a particular training and would return home when the specific task is completed. I did not ask.

Miss Marguiles told us about some of the situations she and her fellow social workers sometimes have to handle. These deal mostly with *the parents* of retarded children.

One mother whose only child so far is retarded managed reasonably well at home but had been getting progressively depressed over the past 18 months. After persistent psychiatric treatment for depression, it seemed the best course was to have the child placed in an institution. The mother resisted the idea at first, but finally agreed. When she phoned earlier in the day to say that she would like to have the child placed, she collapsed and had to be hospitalized.

Other problem situations related to the placement of a child center on cultural differences among parents. While generalizations cannot be validly made, most parents have tremendous guilt feelings when their child is retarded. Parents seem to feel directly responsible for the misfortune.

Many an Italian parent often feels nothing is beyond the doctor's ability. One often gets the impression that they expect the doctor to open the child's brain and put some grey matter in the skull.

Many a WASP parent accepts a mentally retarded child with religious fatality. *God gave me this to chastise me or as a cross to bear to make me a better person.*

Jewish parents tend to see the retarded child as a personal insult. Perhaps because of the high expectations for the child in terms of personal achievements, a mentally retarded child is likely to be ego-deflating to the parents.

There are other problem areas. A mentally retarded child has citizenship problems if the parents are new immigrants. If the diagnosis is made in the home country, the child cannot come to Canada. If the diagnosis is not made until the child arrives with the parents in Canada, it is difficult, if not impossible to get a landed immigrant status for the child. This is an added problem for such newly arrived parents, especially the Eastern European ethnic groups who have to cope with a retarded child, a new environment, occupational, financial and language problems.

One can safely say that no matter what the background of the parents, the social and psychological problems of retardation have many challenging aspects.

Other interesting questions arose. Parents cannot decide that they simply do not want their retarded Mongoloid child and bring him to the Centre for help in placement. Yet even with the normal atomic family (father works, mother works, the couple lives in an apartment and have one child), the first six months or so of a normal child demands almost round-the-clock care. This puts tremendous drain on the parents' physical and emotional resources. If the child is mentally retarded or has a similar problem of early infantile autism, this could make such parents a physical or mental wreck. Is it fair to impose this child on that family? There is no clear-cut answer. Moreover, if the situation were that severe, the child would probably be institutionalized. We saw an 8-year-old girl who was severely retarded, withdrawn, who does not talk or respond to stimuli, but screams and shrieks periodically. Her parents simply do not want her at home. She is still at No. 2 Surrey Place, awaiting placement in an appropriate institution.

We left the second floor and passed through door after door into slowly tunnelling corridors with crossbars. It was a somewhat strange feeling. Miss Marguiles commented: *This is not a jail although it looks like one.* Then I remembered that No. 2 Surrey Place used to be the Toronto Psychiatric Hospital. The architecture of the Clarke's Institute of Psychiatry is a welcome alternative.

We finally made our way to the first floor where there is a classroom. We were met at the door by Harold Palmer, one of the teachers.

The classroom is, in my experience, unique. It has brightly coloured walls, a blackboard, a television set, a refrigerator, stoves and a dinette set, all in a room that measures about 40 feet by 20 feet. The surprised look on our faces must have betrayed us. *We not only try to make the children attempt to read, write and draw, we also try to give them concepts like kitchen, refrigerator, plates, forks and knives.* Harold said apologetically. The walls are gaudily decorated with colour photographs carrying bold captions: BOY, GIRLS, DOG, PINK MEANS SHOWER, BLUE MEANS WATER, etc.

In answer to the question of what it takes to be a teacher to such children, Mr. Palmer said that the regular teacher training plus special courses in instructing retarded and otherwise handicapped children were necessary. He did not mention patience and unusual optimism, but Sam Silverberg did as an inaudible aside. Sam was right.

The teachers get encouragement and satisfaction when an 8-year-old child finally recognises his own name in print after six months of trying hard. It seems that people like Mr. Palmer never give up trying. *You sort of goad them along every inch of the way. You keep reminding them that to make a '3' you have to go out of the window, come in through the door, go out again through the door, and come back inside through the window on the other side of the door.*

Some of the efforts of the children are proudly displayed on the walls.

We heard a lot more about some of the other activities of the Centre. Most of its services are provided on an out-patient or day care basis. The children we saw in the centre are there for a short-term stay, four to six weeks at the most. Most of them are in transit, as it were, to some other hopefully more permanent situation – back to their families or to special institutions for the retarded, such as the one in Orillia. The centre is involved in multidisciplinary research in the various aspects of mental retardation and is active in research in the field of Cytogenetics.

We thanked our hosts for a quite informative tour; we had seen only a few facets of the activities in the Centre. I went upstairs again to room 309 to see a friend, Diana Lang, a social worker in the Centre. Last summer, I had asked Diana to suggest suitable reading material on the social problems of mental retardation especially with reference to parents and siblings of the affected child. Diana had finished her work for the day and was leaving for home. The literature is probably not necessary. There was enough to think about from the afternoon's visit to the centre.

No. 2 Surrey Place probably has its problems and shortcomings that we did not get to hear about. It seems to represent a genuine attempt to cope with a very real social problem. No matter how little the success, it is significant. Of the other ten projects listed on the Social Paediatrics programme, four other institutions sound particularly interesting to visit. These are the Ontario Crippled Children's Centre, the Canadian Institute for the Blind, the Beverley Street School (for Culturally deprived children), and the Juvenile Court. Spending an afternoon in each of these places should be quite informative.

WEBSTER PRIZE

The winner of the Webster Prize this year is Michael Pascoe 7T1 for his article **Old Age and Dying** (Geriatrics issue, December 1970).

Each year the Academy of Medicine presents $50 cash from a sum bequeathed to the Academy many years ago by the widow of Dr. J. D. Webster. The prize is awarded to the undergraduate author of the article judged the best in the **Journal** for the year.

We congratulate Michael and thank the Academy for continued interest in undergraduate journalistic and scholastic efforts.

T.O.

JOURNAL 1971-72

The following themes have been chosen for this "Journal" in 1971-72.

November 1971	Endocrinology
December 1971	Respiratory Medicine
January 1972	Gastroenterology
February 1972	Biochemistry & Genetics
March 1972	Engineering in Medicine

In addition there will be a series of articles on Medical Education with particular emphasis on the new curriculum at the University of Toronto.

Members of the medical classes of 7T2, 7T3, 7T4 and 7T5 are invited to write articles for publication.

Interested individuals should contact the Journal Office, Room 2141, Medical Sciences Building.

F. GENTILI
Editor 1971-72

Grand Round Quiz: Paediatrics

G. Hahn, M.D.
M. Shipman, M.D.
Residents, The Hospital for Sick Children

1. *A 14-month-old child is brought to your office with a two-month history of running nose, cough and intermittent wheezing. Various antibiotics have failed. On examination he is afebrile, has nasal discharge, intercostal indrawing of mild degree, prolonged expirations, but no signs of pneumonia.*

Your management in order of preference is:

1. antibiotic
2. rectal aminophylline
3. decongestant
4. tedral
5. culture of nasopharynx

2. *The same child returns to Emergency at 16 months in distress: tachypnea (70/m), cyanosis, expiratory wheezing, "silent chest" i.e. decreased air entry on auscultation. He has been given Adrenaline, but is not markedly improved when you see him two hours later.*

Your management in order of preference is:

1. Tedral
2. Correct respiratory acidosis
3. Steroid
4. Determination of Arterial blood gases
5. Croupette with oxygen
6. I.V. fluids
7. Allergy skin testing
8. Insuprel inhalations
9. Aminophylline

3. *A three-month-old girl is admitted with pyuria. A bagged urine culture shows:* Proteus *50,000 colonies,* Klebsiella *36,000 colonies, and* E. coli *100,000 colonies.*

What is the next step?

1. Repeat bagged urine for culture
2. Vulvar wash, bagged urine for culture
3. Mid stream urine for culture
4. Catheter urine
5. Suprapubic bladder aspiration for culture.

4. *The cerebral palsy child with the best I.Q. is likely to be:*

1. spastic
2. athetoid
3. ataxic
4. hypotonic

5. *The upright posture obtained with the use of an orthotic device is desirable in the Spina Bifida child. The reasons in order of importance are:*

1. mobility
2. bone growth
3. urinary drainage
4. energy conservation
5. emotional factors.

6. *Central and nephrogenic diabetes insipidus are best differentiated by the use of (one answer only):*

1. skull X-ray
2. EEG
3. water deprivation test
4. renal function studies
5. hypertonic saline infusion
6. Pitressin response test

7. *True or False:*

1. The Guthrie Test for PKU is specific for urinary metabolites of phenylalanine
2. The Guthrie Test can only be done in the Newborn Period
3. Every infant with a positive Guthrie test must be started on a low phenylalanine diet
4. Serum phenylalanine levels must be kept extremely low to prevent mental retardation.

8. *In congenital Cytomegaloverus Inclusion Disease, the most helpful diagnostic aid is:*

1. maternal history during pregnancy of signs and symptoms of infection
2. high IgM titre in infant
3. clinical syndrome in newborn
4. complement fixation test
5. isolation of virus by showing its cytopathogenic effect

9. *A four-week-old infant has had several days of diarrhea treated unsuccessfully with antibiotic and then successfully with nothing but 5% G/W p.o. He is started on skim milk feeds and diarrhea immediately resumes. The most likely causes are:*

1. infection has persisted
2. milk protein intolerance
3. disaccharide intolerance
4. formula should be thickened
5. super-infection with Candida albicans

10. *In the following conditions one may see fat globules in the stool:*

1. Celiac Disease
2. Cystic Fibrosis
3. Administration of fat soluble vitamins
4. Use of fatty creams in perianal area
5. Cyclic Neutropenia, Growth Retardation, Pancreatic Exocrine Deficiency
6. Abetalipoproteinemia

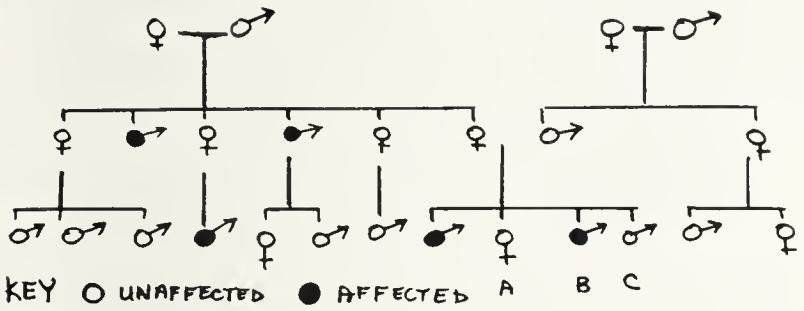
11. *The parents of three children all suffering with cystic fibrosis are expecting their fourth child. What chance has the new baby of being unaffected?*

1. 25%
2. 50%
3. 75%
4. 100%

12. *As a general rule, inborn errors of metabolism, in which specific enzyme deficiencies have been established are manifest only in subjects homozygous (or hemizygous in x linked conditions) for the mutant gene.*

1. True
2. False

13. *See the attached pedigree. By what method is the condition transmitted in this family? What is the chance that the following would be affected?*



1. son of A	0%	25%	50%	100%
2. son of B	0%	25%	50%	100%
3. son of C	0%	25%	50%	100%

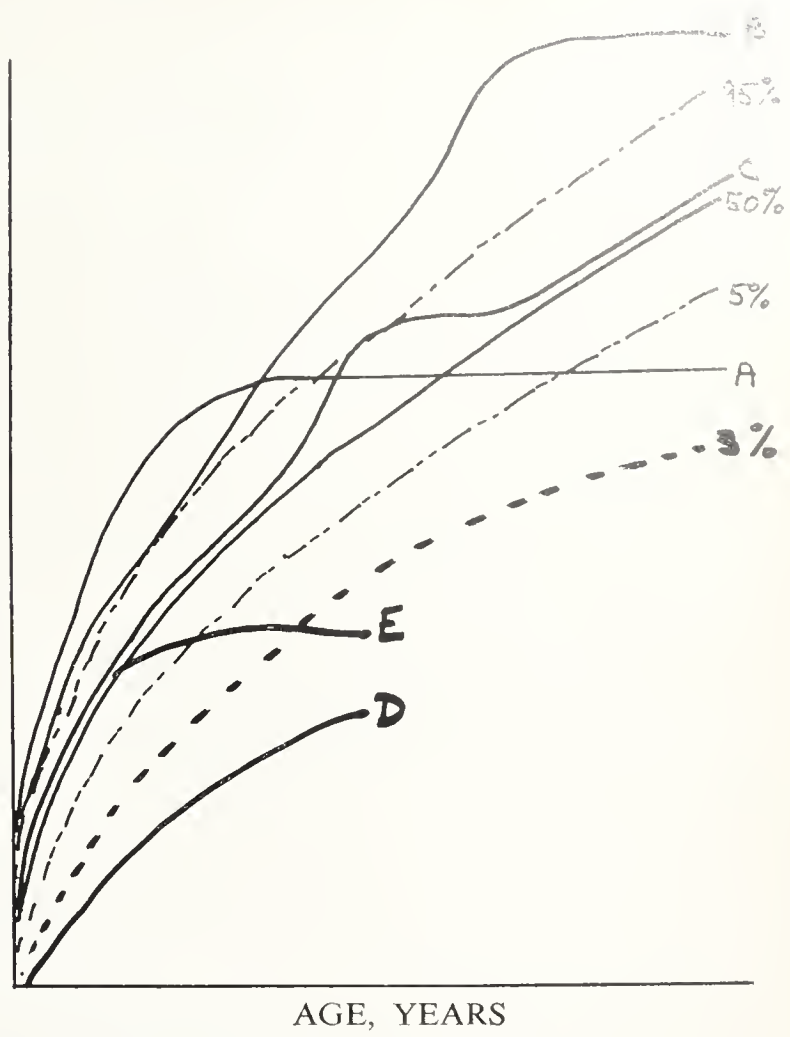
14 $A \pm B \xrightarrow{E+C} X \pm Y$

The above equation depicts a metabolic process in the body. A is converted into X under the influence of an enzyme E acting in the presence of coenzyme C. (B is a second substrate required only in some metabolic processes and Y is a byproduct, again only found in some and not all reactions.)

In an inborn error of metabolism deficiency of a specific enzyme activity results in a block in the conversion of A to X. In the following inborn errors of metabolism name the compounds A and X and the defective enzyme E. If the coenzyme C is of therapeutic importance name that also.

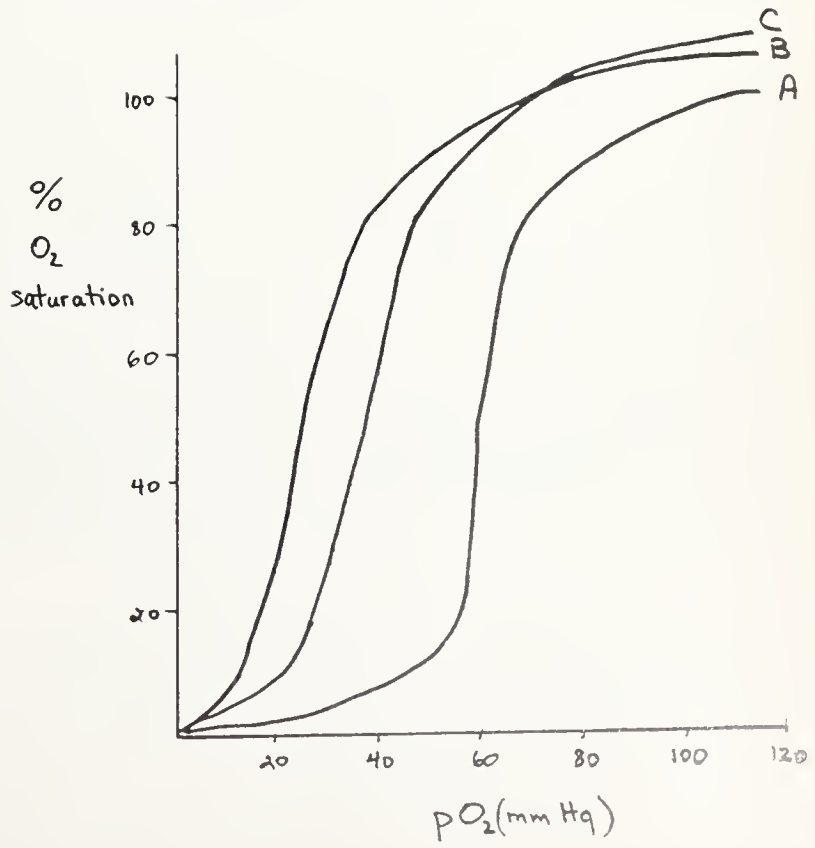
	A	X	E	C
Favism				
Homocystinuria				
Galactosemia				
Phenylketonuria				
Type I Glycogen Storage Disease (Von Gierke's)				
Cystathionuria				
Intestinal Sucrose Intolerance				

15. *Graphs A through E depict growth patterns seen in various diseases. Select the appropriate graph for the untreated conditions listed below.*



1. Coeliac Disease
2. Functioning Chromophobe Pituitary tumour
3. Riley-Day (Dysautonomia)
4. Adreno-genital syndrome
5. True precocious puberty
6. Cystic Fibrosis

16.(a) *One of the curves correctly depicts the dissociation of oxyhaemoglobin at Hb14 Gms%, pH of 7.35 and pCO_2 of 40 mm. of mercury.*



Which one is it?

(b) *Which of the following shift the curve to the right (i.e. at a given pO_2 cause more oxygen to be released).*

1. Acidosis
2. Alkalosis
3. Anaemia
4. Fetal (as opposed to adult haemoglobin)
5. Rise in pCO_2
6. Rise in temperature
7. Presence of Alcohol
8. Inhalation of cigarette smoke

(c) *Which, if any, of the above are associated with a shift of the curve to the left?*

17. *A four-year-old child has suffered resection of the whole ileum following a volvulus. A year later he is admitted for investigation and management of malnutrition.*

(a) *He has steatorrhea – which of the following is likely to be the most important cause of poor fat absorbtion:*

1. Lack of absorbing surface
2. Lack of pancreatic enzyme
3. Stasis
4. Lack of bile salts

(b) *He is almost certain to be depleted in substances whose absorbtion is normally restricted to the ileum. Which of the following are likely to require parenteral supplementation.*

1. Folic Acid
2. Vitamin B_{12}
3. Vitamin D
4. Calcium
5. Iron
6. Copper
7. Magnesium
8. Pyridoxine

Answers

1. 5, 3, 4.
Antibiotic would unlikely be helpful as the aetiology is most likely viral. Rectal aminophylline is to be discouraged as absorption is irregular, and expulsion can occur.

2. 4, 6, 5, 2, 9, 3.
Tedral is ineffective in acute stage of severe bronchospasm.

Skin testing is not advised until after 2 years of age. Isuprel is not to be used if adrenaline has been used within the preceding four hours. Cardiac arrest has been known to occur.

3. 2.
The fact that 3 organisms were grown is suggestive of contamination. The vulvar area requires careful antiseptic preparation before application of a bag. The bag should be applied after a feed, and should be promptly removed when a specimen is obtained.

A midstream urine is desirable but difficult to obtain. The best time is again after a feed. Tapping of the suprapubic area with a finger helps.

Catheterization should be reserved for special procedures.

Suprapubic bladder aspiration has dangers of osteitis, hematoma, and perforation of a viscus. It is sometimes of help in sterile pyuria to decide whether WBC's are of urethral - vulvar origin or not.

4. 2.

5. 5, 2, 3.
1 and 4 are not factors because the child is more mobile, and uses less energy in a wheelchair.

6. 6.
Several days of I.M. pitressin in oil (0.25 c.c.) will allow reduction in urine volume and urine concentration in the former but not in the latter condition in which renal tubules are unresponsive to endogenous or exogenous vasopressin.

7. 1. False. It is a bioassay of serum phenylalanine.
2. False.
3. False. Ratio of false positive to true positive is high.
4. False. If levels of this essential amino acid are too low (less than 3 mg %) then several disturbances in growth occur. It is now thought best to keep the level at 15 mg % or slightly less.

8. 4.

9. 3, 2.

10. 1, possibly; 2, 4 and 5 definitely.

11. 75%

12. True

13. X-linked recessive
1. 25%
2. 0%
3. 0%

	A	X	E	C
14. Favism	Gluc-6-P	6 Phospho-gluconolactose	G6Pde-hyroxylase	
Homocystinuria	Homocysteine (serine)	cystathionine	cystathionine synthetase	P.P.
Galactosaemia	Galactose-1-Phosphate	U.D.P.-Galactose	Ga1-1-P-Uridyl Transferase	
Phenylketonuria	Phenylalanine	Tyrosine	Phenylalanine Hydroxylase	(?F.A.)
Type I Glycogen Storage Disease (Von Gierke's)	Gluc-6-P	Glucose	G-6-P ase	
Cystathionuria	Cystathinonine	Cysteine	Cystathinoninase	P.P.
Intestinal Sucrose Intolerance	Sucrose	Glucose Fructose	Sucrase	

(Note: P.P. = pyridoxyl phosphate)

15. 1. E
2. B
3. D
4. A
5. C
6. D

16. a) 1. B
b) 1, 5, 6, 8.
c) 2, 3, 4.

17. a) 4
b) 2, 7.

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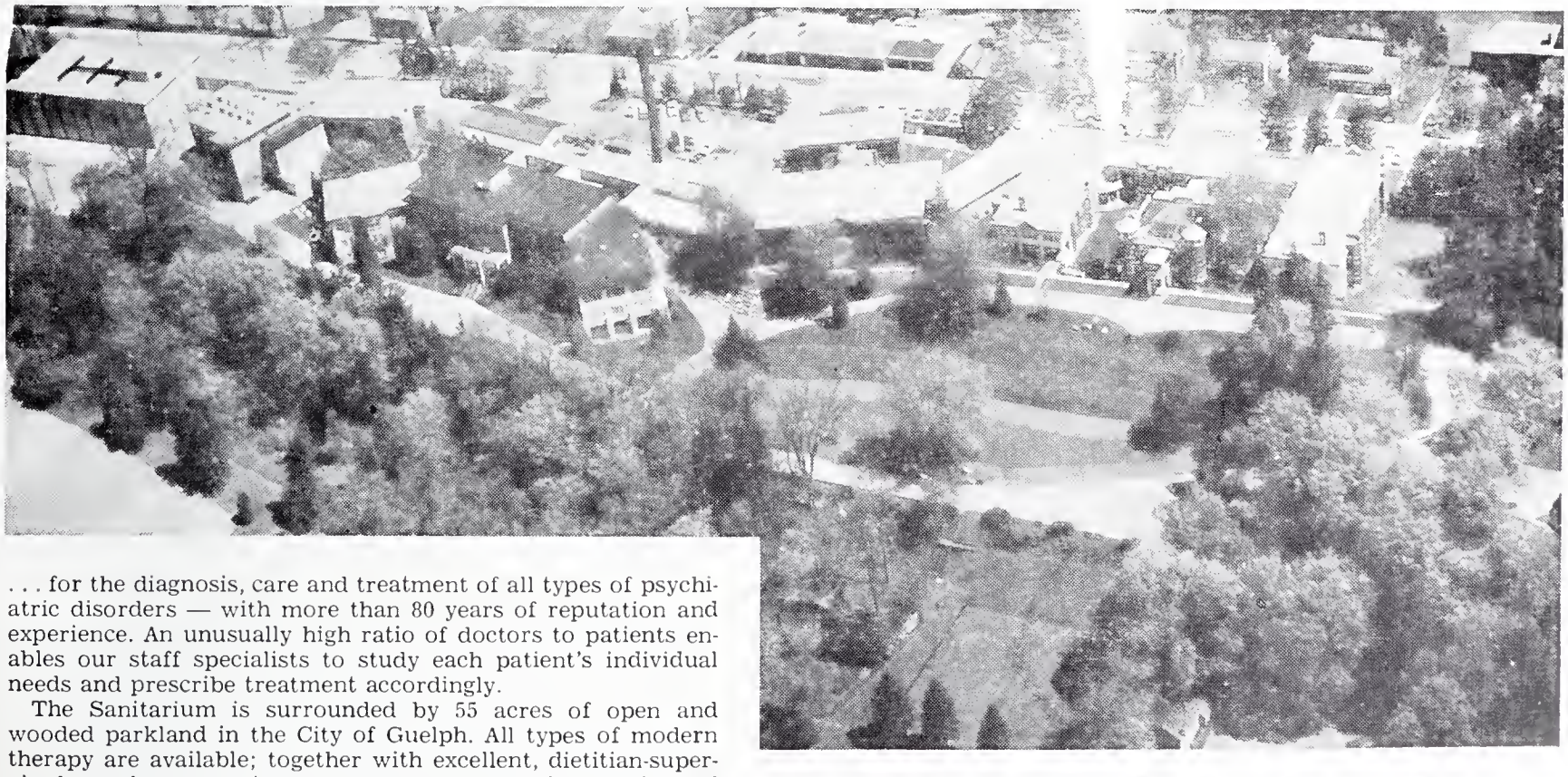

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